TERATOMA OF THE NECK

REPORT OF TWO CASES AND REVIEW OF THE LITERATURE

BY

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Several difficulties were experienced in the diagnosis and management of the two cases which we are reporting. This was due to a lack of detailed knowledge of these tumours, which in turn was due to the absence in the literature of any adequate description of them and especially of their clinical features and treatment; this despite the three reviews which have so far been published (Saphir, 1929; Pusch and Nelson, 1935; Bale, 1950).

It is the object of this paper, therefore, to present a description, gleaned from reviewing the literature and from our own experience, of these tumours, their clinical manifestations and management. The

Definition

Teratomas of the neck are benign cystic, semi-cystic or solid tumours derived from the three germ layers. They are most commonly present in the foetus *in utero*, are sometimes associated with hydramnios and occasionally cause obstruction to labour. The infant may be full-term or premature; it may be stillborn, but is frequently alive at birth. The tumours often cause interference with respiration or swallowing at birth. Sometimes the tumours only become manifest later in infancy or childhood and rarely they may appear in adult life, in which case they are usually malignant.

TABLE 1
SUMMARY OF FINDINGS REVIEWED AND REPORTED BY SAPHIR AND BALE

Author	Sex			Thyroid Tissue			Brain Tissue			Thyroid Gland			'Displacement' of Thyroid Gland				Hydramnios			Operation performed			Thyroid Arteries Entering Tumour		
	M.	F.	No Ref.	Yes	No	No Ref.	Yes	No	No Ref.	Pre-sent	Ab-sent	No Ref.	Yes	No	Doubt-ful	No Ref.	Yes	No	No Ref.	Yes	No	No Ref.	Def-in-ite	Doubt-ful	No Ref.
Saphir (1929): 29 cases re-viewed; 1 case reported	10	13	7	15	11	4	24	0	6	19	4	7	17	0	6	7	4	12	14	17	13	0	2	3	25
Bale (1950): 26 cases re-viewed; 4 cases reported	10	9	11	7	18	5	23	2	5	Not mentioned			19	1	2	8	9	8	13	13	17	0	3	0	27

two case reports illustrate the problems which can arise in dealing with these cases and add to the literature, which is not large.

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The tumours, which appear in the newborn infant and cause interference with respiration and swallowing, present as an urgent matter for diagnosis and treatment. This should not be a difficult problem, as will be shown, and the mortality from excision of the tumour is low provided that surgery

TABLE 1
 SUMMARY OF CASES NOT PREVIOUSLY REPORTED

No.	Author and Year	Age	Sex	Race	Thyroid Tissue	Brain Tissue
1	Marescot (1945)	15 yr.	N.R.	N.R.	'Follicles with colloid'	N.R.
2	Lantuéjoul and Truffert (1946)	Birth	F.	N.R.	Yes	N.R.
3	Hellmuth (1950)	Birth	M.	N.R.	N.R.	Yes
4	Pozzi (1950)	Birth	M.	N.R.	Yes; near capsule	Yes
5	White and Gosselin (1952)	Birth	M.	White	Yes; near capsule also	Yes
6	McGoon (1952) Case 1	Birth	F.	Negress	N.R.	Yes
7	Case 2	Birth 34 wk prem.	M.	Negro	N.R.	Yes
8	Case 3	? 8 mth.	M.	Negro	'Gland structures in capsule'	Yes
9	Case 4	Birth	M.	N.R.	N.R.	Yes
10	Salviati and Savegnago (1952)	Birth	N.R.	N.R.	N.R.	Yes
11	Perkins and Pautler, (1953)	Birth	F.	N.R.	Yes; in capsule also	Yes
12	Otken (1953)	Birth	M.	N.R.	Yes; beneath capsule	Yes
13	Hinds, Seybold and Walker (1954)	Birth	N.R.	Negro	Yes	Yes
14	Buckwalter and Layton (1954)	28 yr.	F.	N.R.	N.R.	Yes
15	Kresse (1954)	Birth	M.	N.R.	N.R.	N.R.
16	Cavallero (1954)	24 yr.	M.	N.R.	Yes	Yes
17	Malaspina and Somaglini (1955)	Birth	M.	N.R.	N.R.	N.R.
18	Kaminek and Tomik (1957)	Birth 27 wk. prem.	F.	N.R.	N.R.	Yes
19	Salas, Esparza, Angulo and Castañeda (1957)	Birth	F.	N.R.	N.R.	Yes
20	Thomas (1957)	Birth	F.	W. African Negress	N.R.	Yes
21	Silberman and Mendelson (1960) Case 1	Birth	M.	White	Yes	No
22	Case 2	Birth	F.	White	Yes	Yes

N.R.

is undertaken before deterioration in the condition of the infant has occurred.

Incidence

Saphir (1929) reviewed 29 cases and added one of his own. Bale (1950) reviewed 26 additional cases and added four of his own (Table 1). We have collected 20 previously unreviewed cases (Table 2) which include a case (Marescot, 1945) mentioned but not included in Bale's review (the second case in Marescot's paper is not included because of lack of histological confirmation of the nature of the tumour), a case omitted by Bale (Lantuéjoul and Truffert, 1946) and 18 other cases. There is therefore a total of 82 cases in the literature

to date, including the two reported in this paper.

These tumours are not quite as rare as might be thought when compared with the number of reported cases of teratoma in other sites, e.g. retroperitoneal teratoma (59 cases up to 1949) or mediastinal teratoma (245 cases up to 1945), but these included dermoids and epidermoids as well as teratomas. (These figures are quoted by Gross, 1953.)

Case Reports

Case 1. A full-term white male infant was admitted to Hope Hospital, Salford as an emergency on April 25, 1957, having been born at home three hours earlier. The mother, aged 28 years, had had two previous normal deliveries. Pregnancy in this case was normal; labour

PREVIOUSLY REVIEWED

Thyroid Gland	Hydramnios	Signs and Symptoms	Treatment and Results
N.R.	N.R.	N.R.	Operation at 36 yr.; no reference to result
Not seen; tumour in position of gland	Yes	Dyspnoea and apnoea	Operation eighth day; no anaesthetic; died post-operatively
N.R.	N.R.	Stridor; cyanosis	Operation at 3 days; post-operative jaundice; well at 2 mth.
Not seen; tumour in position of gland	N.R.	Asphyxia	Died shortly after birth
Not seen	No	Vomiting; dyspnoea	Excision at 3 days; well at 7 mth.
Normal gland	N.R.	Fever, hoarseness and anorexia at 6 mth.; bronchitis and pneumonia	Treatment of pneumonia died 2 wk. later
Right half indistinct	N.R.	Cyanosis; weak respiration	Died 1 hr. after birth
N.R.	N.R.	Difficult respiration and respiratory infection at 8 mth.	Operation and post-operative tracheostomy; well 3 wk. later
N.R.	N.R.	Vomiting; respiratory distress	Caecostomy for imperforate anus; died 2 wk. later
? Left lobe seen	N.R.	Asphyxia; feeding difficulties at birth, then asymptomatic	Operation (local anaesthetic) at 40th day; temporary post-operative paralysis of vocal cord; well thereafter
Absent right lobe	No	None	Excision 6th day; post-operative temporary paralysis of vocal cord; well at 4½ mth.
Attached to superior pole of gland	No	Increase in size of tumour	Enucleation at 3 wk.; well at 6 wk.
Attached to thyroid	N.R.	None	Excision at 3 yr.; uneventful recovery
Involved right lobe and displaced left	N.R.	Ache; increase in size of thyroid	Total thyroidectomy and radical neck dissection; post-operative radiotherapy and ¹³¹ I treatment; died of metastases 1½ yr. later
N.R.	No	Apnoea	Died shortly after birth
N.R.	N.R.	N.R.	Excision at 46 yr.
Tumour in position of gland	N.R.	None	Operation at 7½ mth.; no reference to result
Left lobe absent; tumour adherent to right lobe	Yes	Apnoea and cyanosis	Died ½ hr. after birth
N.R.	N.R.	Cyanosis at birth; dyspnoea; respiratory infection and atelectasis at 6½ wk.	Tracheostomy and cardiac massage at 6½ wks.; died
N.R.	N.R.	Slight stridor at 4 mth.	Enucleation-excision at 4 mth.; well 1 wk. later
Thyroid not seen	No	Dyspnoea and cyanosis at birth	Excision 6 hr. after birth; well at 2 mth.
Normal thyroid gland	No	Rapid respiration; retained secretions; 'moist' chest	Excision at 4 days; died 2 days later; bronchopneumonia

reference

lasted approximately 10 hours and was reported by the midwife in attendance as having been uncomplicated. Both parents were healthy and there were no known familial diseases.

The infant, who was well developed, was extremely ill with very rapid respiration which was 'grunting' in character; there was deep cyanosis and marked rib recession; the heart and lungs were normal. The rest of the general examination was negative.

EXAMINATION OF NECK (Figs. 1 and 2). There was a spherical tumour about 7.5 cm. in diameter situated on the left side of the neck extending from the parotid region above to the clavicle below and from over the mid-line medially well into the posterior triangle. It was hard, without any cystic areas, and its surface was bosselated. The skin over the surface of the tumour

was freely mobile. On its deep surface the tumour appeared to have some attachment in the vicinity of the larynx and thyroid gland but was only loosely attached elsewhere so that the entire tumour was mobile in all directions and hung down over the anterior chest wall because of its weight. The left pinna was displaced upwards and the left half of the body of the mandible appeared to be displaced upwards. The larynx and trachea were obscured by the medial border of the tumour.

The infant was placed in an incubator and given continuous oxygen with some improvement but soon became again deeply cyanosed and limp and appeared to be moribund. Coramine 0.5 ml. and eucortone 0.5 ml. were given intramuscularly and oxygen was given by face mask with marked improvement, but the respiration nevertheless remained difficult and cyanosis persisted.

FIG. 1.

FIG. 2.

FIGS. 1 and 2.—Case 1.
Lateral views of tumour.

A radiograph of the neck (Fig. 3) showed the presence of a soft tissue tumour on the left side of the neck; there was no calcification of the tumour. The trachea was compressed antero-posteriorly; the left half of the mandible showed an absence of the normal angle between body and ramus and these two formed a straight line from the symphysis menti to the base of the skull. The chest was normal.

Tracheostomy was not possible because the tumour overlapped the midline. Excision of the tumour appeared to offer the only possible chance of success. A tentative pre-operative diagnosis of congenital malignant tumour was made.

OPERATION. This was undertaken about three hours after admission under general anaesthesia with endotracheal intubation. A curved skin incision was made, medial and lateral skin flaps were raised and the tumour

was dissected out of its bed with ease because of the well defined capsule and the lack of attachment except in the vicinity of the larynx and thyroid gland, where careful dissection was necessary. It was not possible to ascertain whether the blood supply of the tumour originated in the thyroid arteries or not. No attempt was made to identify the thyroid gland. Haemorrhage during the operation was minimal. The wound was closed with interrupted skin sutures and a Penrose drain was brought out through the wound. The operation was well tolerated.

The drain was removed after 48 hours and the skin sutures on the eighth post-operative day. There were no respiratory difficulties except for stridor, which appeared if the infant was disturbed. Cyanosis did not recur.

The infant was discharged after three and a half weeks

FIG. 3.—Case 1. Lateral radiograph showing soft tissue shadow, compression of trachea and absence of angle of left mandible.

FIG. 3.

FIG. 4.

FIG. 4.—Case 1. Post-operative view of neck.

in hospital (Fig. 4), having gained weight progressively. The wound was well healed, breast feeds were being taken normally and there were no respiratory difficulties.

The infant was asymptomatic and healthy when seen at follow-up two months later.

APPEARANCE OF GROSS SPECIMEN. This was an almost spherical, solid tumour measuring $7.5 \times 6.5 \times 6.5$ cm.

FIG. 5.—Case 1. Photomicrograph showing cartilage in its lower half.

It was greyish-white in colour and the surface was irregular and lobulated. The tumour was well encapsulated.

The cut surface was firm and fibrous with scattered areas of what appeared to be cartilage.

MICROSCOPICAL APPEARANCE (Figs. 5 and 6). Dr. G. J. Crawford (Hope Hospital) reported:

‘The tumour is well circumscribed and in places has a thin fibrous capsule from which fibrous trabeculae run inwards. In several places compressed thyroid tissue is present in the capsule; groups of thyroid acini are also seen in the tumour, strands of epithelial cells and a small island of squamous cells. About half the tumour is composed of fairly cellular fibrous tissue, about one-third of cartilage and the remainder of small areas of myxoid tissue and fat. There is one small area of undifferentiated mesenchyme in which two mitoses were observed. In many of the sections there are scattered foci of haemopoiesis; some of these are associated with fat and thin-walled blood vessels as in bone marrow. The tumour is well supplied with blood vessels, mostly capillaries. No nerve tissue was found. All the tissues in the tumour are either found normally in this area or could be formed from the local mesenchyme. I consider that it is a malformation (hamartoma).’

The tumour was shown to the Children’s Tumour Registry of the University of Manchester whose panel of pathologists kindly examined the sections. Their reports and comments are, with their kind permission, given below.

FIG. 6.—Case 1. Photomicrograph showing thyroid follicles.

Professor A. C. P. Campbell (University of Manchester):

‘Either a teratoma or a mixed tumour of ectopic tracheo-bronchial anlage. I think it is more probably the latter; all the tissue elements present are compatible with a tracheal origin (explaining the squamous epithelium as metaplastic). It appears benign.’

Dr. Agnes R. Macgregor (Royal Hospital for Sick Children, Edinburgh):

‘The tumour is composed of a mesoblastic tissue among which are many masses of cartilage. The mesoblastic tissue varies in character from a moderately cellular collagenous tissue to virtually undifferentiated mesenchyme, in which mitoses are numerous; there are also myxomatous areas. There are atrophied thyroid vesicles at the periphery and epithelial acini, probably of thyroid tissue, appear here and there in the substance of the tumour. No other varieties of tissue are identified.

Inference: It may be a teratoma, but as the epithelial elements appear to be thyroid tissue and all the other elements are mesoblastic, it seems to fit very well the description of the “mixed tumour of the thyroid” mentioned by Willis in *Pathology of Tumours*, 1948, p. 616. The less differentiated parts are histologically malignant.’

Dr. H. Marsden (Royal Manchester Children's Hospital):

'There is loose mesenchymal tissue with spindle cells and numerous islands of cartilage. In addition follicles lined by cuboidal epithelium and containing eosinophilic material are noted. The latter resemble thyroid follicles. The picture is that of a benign teratoid tumour.'

Dr. H. Russell (Christie Hospital, Manchester):

'A defined mass of anomalous mesenchyme in which well-formed cartilage is conspicuous. There appears to be a rim of thyroid tissue closely applied to its periphery. An anomaly of development of the branchial arches.'

R. A. Willis (Prof. Emeritus, University of Leeds):

'A most unusual tumour, composed of moderately cellular vascular mesenchyme with many areas of chondrification, in which some thyroid vesicles and tortuous epithelial strands are incorporated. Thyroid tissue is also present around its well-defined margin. The two possibilities are (I) teratoma, and (II) a chondromatous hamartoma of the thyroid; because of the incorporation of thyroid tissue, I incline to (II). But further sections should be made, in search of other teratomatous tissues. In my opinion the growth is probably benign.'

Even though no uniformity of opinion has been expressed by the panel of pathologists, many of them classified the tumour as a teratoma. For this reason and also because of the extremely close resemblance, both clinically and pathologically, of this tumour to the cases previously reported in the literature as teratoma of the neck, it has been thought quite justifiable to classify it as such.

Case 2. A full-term white female infant was admitted to Memorial Hospital, New York, on June 28, 1949, having been born elsewhere 21 hours previously. The mother, a 28-year-old primipara, had persistent nausea throughout the pregnancy which was otherwise uneventful. Labour lasted 20 hours and the infant was delivered by mid-forceps because of a persistent occipito-posterior position. The weight at birth was 7 lb. 4 oz. The infant had required suction of the mouth and pharynx at birth because of an accumulation of secretions; she had vomited fluids given to her by mouth.

The infant was well developed and was in mild distress from accumulated secretions in the pharynx. Respiration was 44 per minute, heart rate 144 per minute and temperature 99° F. There were numerous rhonchi in the left lung anteriorly. The rest of the general examination did not reveal any other significant findings.

EXAMINATION OF NECK (Figs. 7 and 8). There was a firm, rounded, mobile mass about 14 cm. in diameter situated in the left side of the neck. It extended from the midline of the neck into the posterior triangle; superiorly it extended onto the face and reached the level of the external auditory meatus. Inferiorly, it extended

FIG. 7.—Case 2.
Anterior view of tumour.

FIG. 8.—Case 2.
Lateral view of tumour.

to the root of the neck. The head was displaced to the right. There were a few dilated vessels on the surface of the mass. The anterior part of the mass appeared to be cystic but the rest of the tumour was solid.

INVESTIGATIONS. A blood count and chemical and microscopical examinations of the urine were normal.

A chest radiograph was normal.

A radiograph of the neck (Fig. 9) showed that the tumour contained multiple areas of calcification; the trachea, larynx and oesophagus were normal.

The tumour was aspirated and found to be multilocular; 50 ml. of sero-sanguinous fluid was obtained.

The infant took glucose feeds but vomited frequently; her weight dropped to 6 lb. 6 oz. There were periods of

FIG. 9.—Case 2. Lateral radiograph showing calcification in tumour.

cyanosis and difficulty with respiration, which required frequent suctioning. It was decided that an emergency operation would be necessary. The pre-operative diagnosis was between cystic hygroma, branchial cleft cyst and dermoid cyst.

OPERATION (JULY 1, 1949). Without anaesthesia, an H-shaped incision was made and the tumour was dissected from its bed and removed with its capsule intact. The origin of the blood supply of the tumour was not accurately identified and the thyroid gland was not visualized. The skin was approximated with fine nylon sutures and a Penrose drain was brought out from the wound. The operation was well tolerated.

The infant was placed in a steam tent with oxygen and was given injections of penicillin. She was given tube feeds because she was unable to take oral feeds, but some regurgitation occurred. Thirty-six hours post-operatively her temperature rose and she developed massive consolidation (or collapse) of the left lung and broncho-pneumonia of the right lung. Her condition deteriorated rapidly and she died on the second post-operative day.

AUTOPSY (JULY 4, 1949). This showed that death was due to bilateral severe broncho-pneumonia with abscess formation; aspirated material was present in a few of the bronchi. The thyroid gland, which was grossly normal, was removed and found to be histologically normal.

projected a mass measuring $5.5 \times 5 \times 4$ cm. The lining of the cyst was grey to grey-tan in colour and contained a few yellow-grey plaques up to 3 mm. in diameter. The sectioned surface of the mass was made up of innumerable tiny thin-walled cysts measuring from 1-10 mm. in diameter. These cysts contained clear mucoid material. Scattered throughout the mass were areas of calcification up to 8 mm. in diameter.

FIG. 11.—Case 2. Cut surface of tumour.

MICROSCOPICAL APPEARANCE (Figs. 12-14). The tumour was found to be a benign teratoma which

FIG. 10.—Case 2. Operative specimen. Note excellent capsule.

APPEARANCE OF GROSS SPECIMEN (Figs. 10 and 11). A well-encapsulated, grey-tan rounded tumour measuring $7.5 \times 6.5 \times 5.5$ cm. was found on section to consist of a large cystic cavity containing bloody fluid into which

FIG. 12.—Case 2. Photomicrograph showing choroid plexus.

FIG. 13.—Case 2. Photomicrograph showing cartilage.

FIG. 14.—Case 2. Photomicrograph showing thyroid follicles.

contained cysts lined with squamous, cuboidal or columnar epithelium; other cysts contained papillary projections. Bronchial epithelium, smooth muscle, immature cartilage and embryonic connective tissue were identified. Thyroid follicles and brain tissue (nerve fibres, glia, choroid plexus) were also in evidence.

Discussion

Nomenclature. Attempts have been made in the past to define which tumours should be called teratoma of the thyroid gland and which should be called cervical teratoma.

Saphir (1929) stated that if the thyroid gland was absent the tumour was a thyroid teratoma, but that if the thyroid gland was present the tumour was a teratoma of the neck. These criteria, we think, are basically sound but require further elaboration and clarification.

Bale (1950) disagreed with Saphir's method of classifying the tumours and instead proposed a more complicated nomenclature, viz. (a) teratoma of the thyroid gland if the thyroid arteries supplied the tumour; (b) teratoma of the neck in the region of the thyroid gland if the tumour replaced all or part of the gland; and (c) teratoma of the neck, probably in the region of the thyroid gland, because of the general appearance of the tumour. This classification suffers from several objections. Firstly, the fact that the thyroid arteries supply the tumour cannot be accepted as evidence that the tumour has its origin in the thyroid gland; these arteries supply many structures other than the thyroid gland and a tumour supplied by them may have arisen from any one of these structures, e.g. parathyroids, larynx, soft tissues. Secondly, very few reports in the literature mention the blood supply of the tumour and hence are unclassifiable on Bale's criteria. Thirdly, in carrying out an emergency procedure for a cervical teratoma on a newborn infant, it would be extremely unwise to search for the thyroid arteries if they were not easily visible (this obviously does not apply to autopsy specimens). Finally, the creation of three categories of tumour, as proposed by Bale, appears cumbersome and unnecessary.

We propose that the tumour be referred to as a cervical teratoma unless it fulfils the following criteria, in which case it may be called a true teratoma of the thyroid gland. The teratoma is situated in the position of the thyroid gland and in addition (a) the thyroid gland is present and the tumour occupies a portion of it; or (b) the thyroid gland is only partially present, but the tumour is in direct continuity with it, the two forming one mass; or (c) the thyroid gland is completely absent and the tumour replaces the gland entirely.

When a teratoma is adjacent to, but not in direct continuity with, a thyroid gland which may be only partially present, then the tumour cannot be classified as a true teratoma of the thyroid gland because the tumour may well have arisen outside the gland and given rise to atrophy of the gland by pressure.

The presence or absence in the tumour of thyroid tissue is of no particular value in classification since by their very nature all teratomas are capable of producing a wide variety of tissues, and thyroid tissue is no exception.

Accordingly the following cases which have appeared since 1945 should be referred to as true thyroid teratomas: Cases 2, 4, 12, 14, 17 and 18 (see Table 2), possibly Cases 5 and 13 (see Table 2) and Bale's (1950) first three cases.

Age. The age distribution in 79 of the 82 cases reported to date in which age is mentioned is seen in Fig. 15.

FIG. 15.—Age distribution of 79 cases.

- A = live premature infants
- B = stillborn premature infants
- C = live full-term infants
- D = stillborn full-term infants
- E = birth to 1 month
- F = 1 month to 1 year
- G = 1 year to 15 years
- H = adults

Neither Saphir's (1929) nor Bale's (1950) reviews state whether they refer to the date of appearance of the tumour or the age of the patient when first seen. Nevertheless, it can be presumed that most, if not all, of the cases up to 1 month old and many of those up to 1 year old had the tumour at birth. In those cases which we have collected we have taken care to utilize the age at appearance of the tumour and not the age of the patient when first seen.

In any case, it can be seen from Fig. 15 that the majority of tumours were present at birth although some do not appear to present until later. The tumour is rare after the age of 1 year.

Sex. The sex of the patient was mentioned in 61 cases out of 82 in the literature, of which 30 were females and 31 were males.

Race. Although most of the tumours have appeared in white patients, they have also been reported in Negroes (Saphir, 1929; McGoon, 1952; Hinds, Seybold and Walker, 1954; Thomas, 1957).

Histology. All varieties of tissue from the three germinal layers have been found in these tumours. Particular mention has been made in the past of brain tissue, which has been observed in 47 of 60 cases of Saphir's and Bale's and in 17 of our 22 cases, and also of thyroid tissue, which was present in 22 of Saphir's and Bale's 60 cases and in 11 of our 22 cases (see Tables 1 and 2).

The vast majority of tumours were benign on histological examination but, unfortunately, there has been no long-term follow-up on them. Four definite cases of malignancy have been reported to date. Three were mentioned by Bale (1950). The first was a 9-week-old infant, who died on the day of operation (Pupovac, 1896). The second was a 53-year-old woman, who died of metastatic pulmonary sarcoma from probable malignant proliferation of the teratoma (Lurje, 1908). The third was a 41-year-old female, who died one month post-operatively of widespread metastatic sarcoma, probably originating in the teratoma (Fritzsche, 1920). The fourth case was reported in 1954 by Buckwalter and Layton. Their patient was a 28-year-old female, who died of metastases 15 months after total thyroidectomy and radical neck dissection. The tumour had caused progressive enlargement of one lobe of the thyroid gland for a period of six months prior to surgery. On histological examination it was found to have contained immature neural tissue and muscle as well as cartilage, mesenchyme and gland structures.

Signs and Symptoms. The following remarks are based on a close study of all the cases reported since

1945 (see Table 2) and therefore include Bale's (1950) four cases.

The majority of tumours were oval in shape; an occasional one was spherical. In size, except for Cavallero's (1954) case, in which the tumour was the size of a nut, most of the tumours were between 5 cm. and 12 cm. in their longest axis. When compared with the antero-posterior diameter of a newborn infant's head, which is 10 cm. (approximately), it can be appreciated that these tumours are quite large.

Most of the tumours were situated on one or other side of the neck, usually extending up as far as the mastoid process and body of the mandible and often displacing the lobule of the ear upwards; many tumours reached the zygomatic arch. Posteriorly they usually reached the anterior border of trapezius and anteriorly they often crossed the midline for a short distance. The inferior border of the tumour was most frequently the clavicle but in two cases the tumour entered the mediastinum (Salas, Esparza, Angulo and Castañeda, 1957; Kaminek and Tomik, 1957). In four cases (Lantuéjoul and Truffert, 1946; McGoon's third case, 1952; Pozzi, 1950; Malaspina and Somaglino, 1955) the tumour was situated in the position of the normal thyroid gland and was mistaken for a congenital goitre. The tumour reported by Buckwalter and Layton (1954) was the only one which was palpated as a swelling in a lobe of the thyroid gland; but this tumour occurred in an adult aged 28 years who had a malignant teratoma.

The consistency of the tumour varied. It was cystic, or partly solid and partly cystic, or entirely solid. However, in most instances fluctuant cystic areas were palpable. An occasional case was transilluminated. The surface of the tumour was usually irregular, being described as lobulated, bosselated or loculated, and the borders of the tumour were usually well defined.

In only a few case reports was the mobility of the tumour mentioned. In these cases the tumour was usually quite mobile as was the skin on the surface of the tumour. A few reports mentioned that there were collateral veins on the surface of the tumour.

The cases which were not stillborn usually had acute symptoms at birth. These were cyanosis, dyspnoea, apnoea and stridor due to tracheal deviation and/or tracheal compression. In a few instances swallowing difficulties occurred. These symptoms often led to a fatal termination if early treatment was not undertaken (McGoon, 1952; Kaminek and Tomik, 1957). Occasionally, the infant was asymptomatic at birth and only several weeks or months later developed serious symptoms (McGoon, 1952; Salas *et al.*, 1957) and in a few

instances the tumour remained entirely asymptomatic (Otken, 1953; Hinds *et al.*, 1954).

The presence and degree of tracheal deviation and obstruction was often demonstrable radiologically. The radiograph also showed calcification of the tumour in a few cases, as occurred in our second case, in McGoon's (1952) fourth case and in Thomas's (1957) case.

Aspiration of the mass was performed in the case described by Lantuéjoul and Truffert (1946). They obtained opalescent fluid but were not able to empty the cyst of its contents, indicating its multilocular nature. Our second case was aspirated and sero-sanguinous fluid was obtained; aspiration also showed that the tumour was multilocular.

Effects of Tumour on Pregnancy and Labour. There have been two cases of hydramnios in the cases reported since 1945 (Lantuéjoul and Truffert, 1946; Kaminek and Tomik, 1957). Before 1945 13 cases had been reported (Table 1), making a total to date of 15 instances in 82 case reports (18%). This coincides well with the high incidence of hydramnios which is known to occur in cases of foetal malformation. Hydramnios is often associated with anomalies such as oesophageal atresia, in which the foetus is unable to swallow the liquor amnii. This is postulated as an aetiological factor in hydramnios (Eastman, 1956). It is possible that the oesophageal obstruction produced by a cervical or thyroid teratoma is in a similar manner responsible for the high incidence of hydramnios in these cases.

The size of the tumour may impede labour and may require the application of forceps (White and Gosselin, 1952; Kresse, 1954; our second case) or caesarean section (Salviati and Savegnago, 1952) or episiotomy (Perkins, Pautler and Winston-Salem, 1953). In the case reported by Salviati and Savegnago (1952) the tumour was stated to have been palpated in the abdomen and per vaginam.

Associated Congenital Anomalies. Two instances of associated congenital anomalies have appeared since 1945: 'chondro-dystrophia foetalis' was present in McGoon's (1952) second case and imperforate anus in his fourth case.

Differential Diagnosis. Some of the diagnoses, which have been considered by various authors pre-operatively, are cystic hygroma, congenital goitre, branchial cyst, lymphangioma, simple multilocular cyst, dermoid cyst, neuroblastoma, parotid tumour and carcinoma of the thyroid gland (in Buckwalter and Layton's case of malignant teratoma in an adult).

The chief tumours which require differentiation are cystic hygroma, congenital goitre and branchial cyst.

In age incidence, sex incidence, site, size and surface of the tumour, teratoma and cystic hygroma are similar. However, a cystic hygroma usually has a limpid consistency and poorly defined borders and is easily transilluminated (Gross, 1953); teratomas are often tense or solid in consistency, have well defined borders, are usually freely mobile and only occasionally can they be transilluminated. Radiography may demonstrate calcification in teratomas but not in hygromas. Finally, hygromas produce few symptoms, except when infected or very large, which is in direct contrast with the respiratory and feeding difficulties commonly seen with teratomas.

In those instances in which the teratoma occupies the position and has the shape of a thyroid gland it must be differentiated from a congenital goitre. However, since congenital goitres are rare outside endemic areas (McQuarrie, 1957), the problem is practically limited to these areas. Radiography of the neck may be of value in demonstrating the presence of calcification in a teratomatous tumour.

The following features of a branchial cyst serve to differentiate it from a teratoma: it is commonest in the third decade and presents as a smooth, globular swelling deep to the anterior border of the sternomastoid muscle, usually at its middle; on aspiration typically it yields a milky fluid containing cholesterol crystals (Aird, 1957).

Prognosis and Treatment. Forty-six patients of all those reported thus far (82) have undergone surgery, and seven died. Of the deaths, three occurred in patients with malignant tumours: one was an operative death, the second was an adult who died of metastases (both mentioned by Bale, 1950) and the third was also an adult who died of metastases (Buckwalter and Layton, 1954). Of the remaining four deaths one was an infant who died of cardiac arrest due to an attempted tracheostomy (Salas *et al.*, 1957) and another was our second case who died of broncho-pneumonia post-operatively. According to Bale (1950), the cause of death was not mentioned in the remaining two cases. The mortality with surgical treatment is thus about 9% if the malignant tumours are not included, an extremely low figure considering that over half the cases were treated before 1930.

Of the 36 cases in which surgery was not undertaken, 14 were stillborn and one was an adult who died of metastases from a malignant tumour (Bale, 1950). The remaining 21 cases were live, newborn

infants, all of whom died without having had any operative treatment. It is difficult to estimate how many of these infants could have been saved by surgery but, to mention a few instances only, the lack of early surgical treatment was most probably responsible for death in two of McGoon's (1952) cases and in Carter's case (mentioned by Bale, 1950).

Thus the significant facts are that the mortality from operation on benign cases at all ages is low; that more than 25% of all patients died soon after birth without having had surgery; that it is more than likely that many of these infants could have been saved by timely operation.

Hence the first principle in the treatment of these tumours in infants is early operation, especially when tracheal and oesophageal obstruction are present. Delay in surgery, when the respiratory passages are partially obstructed, leads to retention of secretions, atelectasis and broncho-pneumonia. When the tumour does not encroach on the trachea or oesophagus surgery may be postponed, but this is not common.

In the pre-operative phase resuscitative measures such as tracheal aspiration, oxygen and stimulants may be required; laying the infant on the side of the tumour may help to relieve some of the compression and traction on the trachea.

The operation should consist of excision of the tumour and its capsule together with redundant skin. General anaesthesia with intubation should be employed; some authors have employed local anaesthesia (Salviati and Savegnago, 1952) or no anaesthesia at all (Lantuéjoul and Truffert, 1946).

In benign cases, the operation itself is relatively simple since the tumour 'shells out' without much difficulty. Care should be taken when the tumour is loosely adherent to the respiratory passages or thyroid gland since temporary vocal cord paralysis has occurred (Salviati and Savegnago, 1952; Perkins and Pautler, 1953). Caution may be required when, occasionally, the tumour enters the mediastinum.

Tracheostomy without excision of the tumour is not usually possible since the tumour obscures the trachea in many cases. However, tracheostomy has been performed after removal of the tumour (Lantuéjoul and Truffert, 1946; McGoon, 1952) in order to establish and maintain a patent airway.

Post-operatively, meticulous care should be taken in maintaining a patent airway; antibiotics should be given and oral feeding may be carefully commenced fairly soon after surgery. When the thyroid gland is absent or is removed by operation replacement therapy with thyroid extract will be required.

It is not proposed to discuss the management of

malignant teratoma in detail. However, wide local excision of the tumour is obviously mandatory if there is to be any prospect of avoiding local recurrence.

Summary

Cervical teratomas have been defined. Twenty-two new cases have been added to the 60 cases previously reviewed. Two personal cases have been described in detail.

Previous attempts to distinguish thyroid from cervical teratoma have been indicated and a new set of criteria proposed. The tumours usually appeared at birth and were equally divided between the sexes. Several cases have been reported in Negroes.

Histologically tissues from all the germinal layers were found including brain and thyroid tissue. Except for four cases of malignancy (three in adults) all the tumours were benign.

The appearance of the tumour has been described in detail and the likelihood of acute obstructive symptoms of the larynx and oesophagus at birth has been indicated. The tumour caused hydramnios in 18% of cases and occasionally impeded labour. Occasionally congenital anomalies were present in other parts of the body.

The chief points in differentiating the tumour from cystic hygroma, congenital goitre and branchial cyst have been indicated.

The mortality with surgery in benign cases was 9%. On the other hand, all cases not treated surgically died. The principles of surgical treatment have been outlined.

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Addendum

Since this paper was written an article has appeared reporting two further cases of teratoma of the neck (Keynes, W. M. (1959). Teratoma of the neck in relation to the thyroid gland. *Brit. J. Surg.*, 46, 466). A summary of tumours reported since 1950 is given, but omits mention of Cases 1-4, 10 and 15-19 (see Table 2). It provides reference to two cases before 1950 which we had omitted (Shattock, S. G. (1882). Congenital tumour of the neck. *Trans. path. Soc. Lond.*, 33, 289; Daniels, D. W. (1928). Congenital tumour of the neck. *Brit. J. Surg.*, 15, 523).