MALIGNANT CAROTID BODY TUMOUR IN THE NECK OF A NEWBORN INFANT*

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The purpose of this paper is to add a fourth case to the three cases of the rare tumours arising in ectopic chemoreceptor cells. From a study of the published reports concerning such tumours, most of them arise from the known organized collection of chemoreceptor cells.

In recent years reports have appeared of a few cases having the same structure as carotid body tumour, but arising away from the carotid vessels and vagus nerve. The present case conforms in its general behaviour to those cases that arise in the neck, but differs in that it began after birth.

Case Report

N.A., aged 3 months, had been admitted to the Alexandria University Hospital complaining of a swelling of the left side of the neck. The swelling had been noticed by the mother at birth and it grew rapidly during the period before admission. On examination, the swelling was lobulated, occupying the whole side of the neck and encroaching on the posterior triangle. The skin over it was very thin, and numerous dilated veins were seen (Fig. 1). It was soft in consistency and slightly mobile from side to side.

Because of impending ulceration, excision was carried out quickly, and it was noticed during the operation that the tumour, in spite of its size, was encapsulated. The carotid sheath with its contents was displaced medially. The carotid vessels and the vagus nerve could be dissected from the tumour and it was demonstrated during the operation that the tumour had no relation to them.

Pathological examination revealed the following: macroscopically, the tumour consisted of lobulated, fleshy tissue, dark pink in colour, and covered by numerous dilated veins. In cut section its encapsulation was shown. The colour was dark pink and the surface was homogeneous.

Microscopically there was a highly cellular lesion, mainly consisting of acini-like collections of poorly-defined cells, with large vesicular nuclei and either clear or granular cytoplasm. These acini were separated from each other by fibrovascular strands. The mitotic activity was low. Included among the cellular areas were dilated vascular spaces mainly of lymphatic type and these ectatic lymphatics were thought to be part of the

host tissue, suggesting a diffuse infiltrating process (Fig. 2).

The histological and the clinical features suggest a diagnosis of carotid body tumour of the infiltrating type.

The excision was followed by a course of radiotherapy.

Six months later the infant developed a local recurrence on the same side of the neck, associated with Horner's syndrome (Fig. 3). Re-excision showed that the recurrence had the same macroscopical and microscopical features as the primary, but mitotic activity and hyperchromatism were more evident (Fig. 4).

While in hospital, three weeks after the second operation, she developed a recurrence in the nape, which was easily removed. She remained free of metastasis till March 1963 when she returned with supraclavicular and submaxillary masses which were deeply seated and fixed. Irradiation therapy was advised and there was some response. The child is in good health now.

Discussion

Chest radiographs during the whole illness did not reveal any secondaries. This case represents a very rare tumour which had the same structure as carotid body tumours in spite of its being seated away from the carotid vessels. Recently different authors have stated that carotid body tumours may arise in sites away from the known organized collections of chemoreceptor tissue as the carotid body, aortic body, pulmonary body, glomus jugulare and ganglia nodosum of the vagus nerve. Other glomera had been described in or close to the walls of the large arteries of the neck, in the retroperitoneal tissues and in the soft tissues surrounding the femoral vessels (Smetana and Scott, 1951). The function of these ectopic glomera is not understood, but due to similarity of their structure with the carotid body they may have the same function. They may play a part in the haemodynamics of restricted areas but so far the evidence is inconclusive.

Chemoreceptor tumours arising in ectopic chemoreceptor tissue in the neck are rare. Only three cases have been reported: one by Marcuse and Chamberlin (1956), another by Ellis and Winston (1957) and the third by Dallachy and Simpson (1960). These three tumours arose in the side of the neck and were away from the carotid vessels and the vagus nerve.

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FIG. 1.

FIG. 2.

FIG. 1.—The condition of the child when first seen.

FIG. 2.—Microscopical section showing the highly cellular lesion, mainly consisting of acini-like collections of poorly-defined cells, separated from each other by fibrovascular strands. ($\times 288$.)

Single cases of tumours of similar structure have been reported in each of the following sites: trachea (McCall and Karam, 1958), larynx (Andrews, 1955), chest wall (Shaw and Kennedy, 1956) and ciliary ganglion (Fisher and Hazard, 1952). Similar tumours may arise alongside the arteries of the lower

limbs, trunk and neck, and these are prone to give metastases in the liver and the lungs (Smetana and Scott, 1951; Randall and Walter, 1954; Block, Dockerty and Waugh, 1955).

The three tumours of the carotid body type that occurred in the neck outside the carotid vessels and

FIG. 3.

FIG. 4.

FIG. 3.—Recurrence of the growth after excision, associated with Horner's syndrome.

FIG. 4.—Microscopical section of recurrence showing marked mitotic activity and hyperchromatism. ($\times 288$.)

vagal ganglia were smaller than those arising in the organized collections of chemoreceptor tissue. Moreover, they were still apparently encapsulated, grey or pale brown in colour and moderately vascular. The behaviour of these tumours arising in ectopic chemoreceptor tissue is different from those of the carotid body. Carotid body tumours grow slowly, and evidence of malignancy is considered by some authors as a bar to diagnosis (Phelps, Case and Snyder, 1937). According to Romanski (1954) there are records of carotid body tumours with distant metastases, secondary growths in the cervical lymph nodes, sternomastoid, bones, viscera due to direct invasion, lymphatic permeation or blood spread. The commonest overt malignant manifestation is local recurrences following local excision. The high percentage (15-20%) of local recurrence reported by Romanski (1954) was disputed by LeCompte (1951) who found no evidence of malignant disease in 17 cases examined histologically. He observed considerable variations in the number and size of the cells, but he declared that similar changes were seen in benign tumours as in parathyroid adenoma. Willis (1948) stated that with few and doubtful exceptions, tumours of the carotid body were benign, grew slowly, remained sharply circumscribed and produced no metastases.

On the other hand, the structure of chemoreceptor tumours arising in ectopic chemoreceptor tissues found in the muscles of the neck and limbs is different (Smetana and Scott, 1951). They are generally larger, the cells are usually found in well-differentiated acini, and secondary deposits are found in the majority of the cases. Although the cells of these tumours are not less differentiated in their behaviour, malignant deposits are of low activity relatively. The main features encountered in the present cases agree with those reported by Smetana and Scott (1951).

As regards the diagnosis none of the tumours reported before had been diagnosed before operation. Most of the cases had been removed to reach a diagnosis as well as to remove the mass. Clinically the main features of this case are: (1) the appearance of the tumour at birth; this has not been reported before. (2) The rapidity of growth during the first three months. (3) Evidence of malignancy manifested by the presence of Horner's syndrome on the same side, due to invasion of the sympathetic nerves and the recurrence after local removal. (4) The early appearance of the recurrences, which were limited to the area near the operation and on the same side, suggesting an implantation process as a possible mode of spread. (5) Marked radiosensitivity of the recurrences to radiotherapy.

The youngest age reported for carotid body tumour was by Wosnesseki (Gordon-Taylor, 1940) in a child aged 6 months, another was at the age of 5 years by Green (1908). Not a single case of chemoreceptor tumour at birth has been reported.

Evidence of malignancy is quite clear in this case, as noted by the rapidity of growth, recurrence after excision, presence of Horner's syndrome and, histologically, by the presence of mitotic figures.

It is agreed that the best treatment is complete excision. The only obstacle to removal in the neck is the dense fibrosis surrounding and enclosing the vessels and nearby structures.

Post-operative radiotherapy was employed in the three reported cases. Most cases showed response to radiation in the form of growth restraint and reduction in vascularity.

Summary

A rare case (only four have been recorded so far) of chemoreceptor tumour is reported. It originated from cervical ectopic chemoreceptor tissue situated away from the carotid vessels and vagus nerve. It had manifested itself at birth. The rapidity of growth, the recurrence after excision and the presence of Horner's syndrome all testified to its malignant nature. It is the first reported case occurring at birth. Radiotherapy was successful in the treatment of the secondaries.

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