MALIGNANT NON-CHROMAFFIN PARAGANGLIOMA OF THE THIGH

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Within recent years it has become recognized that some tumours arising in skeletal muscle have a close resemblance histologically to tumours of non-chromaffin paraganglia such as the carotid body and glomus jugulare (Smetana and Scott, 1951). Such tumours are rare and have previously been designated granular cell myoblastomata or, more recently, alveolar soft-part sarcomas (Christopherson, Foote and Stewart, 1952). They occur mainly in children and young adults and are often malignant.

The case recorded here is that of a boy who developed a tumour of the left rectus femoris muscle, which recurred after local excision and subsequently gave rise to widespread metastases from which he died a year later.

Case Report

J.T., a boy aged 6 years, attended the Royal Hospital for Sick Children, Edinburgh, in June, 1950, with the complaint of a swelling in the front of the left thigh for three weeks. It had been gradually increasing in size but had given rise to no symptoms.

The swelling was excised locally. He remained well for eight months but then complained of frontal headaches and vomiting. He was found to have bilateral papilloedema and raised intracranial pressure due to a space-occupying lesion in the posterior part of the left cerebral hemisphere. Left occipital craniotomy and decompression were performed. He gradually deteriorated, with herniation of the brain through the craniotomy and paraplegia as a result of spinal metastases. The tumour recurred in the front of the left thigh and there was lymphatic spread to the left inguinal nodes.

Morbid Anatomy

The primary tumour in the left rectus femoris muscle measured 7 x 3 x 3 cm. It was oval, fleshy and generally white with several pale pink areas (Fig. 1). It was embedded in muscle and its margin was well defined except in one region where it appeared to be infiltrating.

At necropsy a year later, the tumour had recurred locally and formed a large, fleshy mass, 15 x 5 x 4 cm., adherent to the femoral periosteum. The left inguinal lymph nodes were enlarged and replaced by tumour. Secondary deposits were present in the liver, lungs, vertebral column and spinal cord, brain (Fig. 2), skull, palate and the medulla of the right adrenal gland. In all these situations except the brain the metastatic deposits were similar macroscopically to the original tumour in the thigh. The large neoplastic mass in the brain differed in that...
eosinophilic cytoplasm. The nuclei were large, variable in position and usually vesicular. Many contained prominent nucleoli. Mitotic figures were scanty. A striking feature in several areas was the production of a pseudo-acinar structure by degeneration of the central cells in the tumour masses (Fig. 6). Invasion of veins was observed in the primary tumour (Fig. 6).

Discussion

Tumours of this type have usually been classified as granular cell myoblastomata because of their relationship to skeletal muscle and granularity of the cytoplasm of the neoplastic cells. They can be readily identified by their distinctive histological pattern in the illustrations of several cases reported as granular cell myoblastomata, e.g., Case 6 of Klemperer (1934), Case 4 of Horn and Stout (1943) and the case of Schwidde, Meyers and Sweeney (1951). Horn and Stout noted the striking pseudo alveolar structure of this group and used the term ‘organoid’ granular cell myoblastoma to describe them. Christopherson et al. (1952) reported 12 widespread necrosis and haemorrhage had occurred. All epithelial tissues were examined with particular care to exclude a possible primary growth other than that in the thigh muscle.

Histology

Both primary and secondary tumours showed an identical histological pattern which is very characteristic and striking. The tumour cells were arranged in discrete groups which were enclosed by delicate, but usually well defined collagen fibres (Fig. 3). In many areas, these cell groups were separated from each other by vascular channels the walls of which were formed by the limiting collagenous membranes with an inner layer of extremely flattened endothelial cells (Figs. 4 and 5). These vascular spaces sometimes contained blood (Fig. 4), but were more often empty (Fig. 5). In the secondary deposit in the brain they were very large and engorged with blood. This delicate layer of endothelium was sometimes extremely thin and in places deficient (Fig. 5) so that tumour cells bulged into the lumina.

The neoplastic cells were large and polyhedral with indistinct cell boundaries and coarsely granular

Fig. 4.—Delicate membranes of connective tissue enclose the groups of tumour cells. Several vascular channels containing red blood cells also present. H. & E. × 300.
tumours of this type, mainly in young adults and arising in skeletal muscle; 50% of them metastasized. They drew attention to their resemblance to paragangliomata but knew of no identification of any paraganglionic tissue in the extremities. They termed them alveolar soft-part sarcomas.

Smetana and Scott (1951) collected 14 examples of this tumour and showed that there was a close similarity to tumours of the carotid body and glomus jugulare. They considered they were paragangliomata and to support this view provided illustrations in their paper of structures removed from the vicinity of the large femoral vessels in Hunter’s canal which they interpreted as normal paraganglionic tissue. Since then further examples have been described (Randall and Walter, 1954; Hicks and Leitch, 1955). Willis (1953) reported that he had seen five such tumours and was of the opinion that they were paragangliomata.

We agree with Smetana and Scott that these tumours are most probably non-chromaffin paragangliomata because of their striking resemblance to tumours of undoubted paraganglia, but conclusive proof of their origin must await confirmation of the presence of normal paraganglia in the limbs. We have attempted, without success so far, to identify such structures in the lower limbs of children and adults at necropsy.

There seems little doubt, however, that this tumour is an entity whatever its tissue of origin. The histological pattern has been reproduced fairly uniformly in all the examples so far published. It appears to arise within or in close proximity to skeletal muscle, usually of the limbs, and affects mainly young adults or children. Many metastasize though they may appear histologically benign. Invasion of veins in the primary growth, as was demonstrated in this case, may indicate the future behaviour of these tumours.

Summary

A malignant tumour arising in the rectus femoris muscle of a young boy is described. It is considered to be a non-chromaffin paraganglioma.

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REFERENCES


