ORBITAL MALIGNANT NON-CHROMAFFIN PARAGANGLIOMA*† ALVEOLAR SOFT TISSUE SARCOMA

BY

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MALIGNANT non-chromaffin paraganglioma, or what has been designated "alveolar soft tissue sarcoma", was described as a distinct entity by Christopherson, Foote, and Stewart (1952). Most of the reported cases are from such sites as the thighs and the retroperitoneum (Stein, 1956; Pack and Ariel, 1958; Karnauchow and Magner, 1963; Vakil and Sirsat, 1963; Bhargava, Bhargava, Jeevandra Kumar, and Srikantiah, 1965; Indira, Gangadharam, Ramachandra Rao, and Reddy, 1965). Orbital cases are very few (Hogan and Zimmerman, 1962).

A girl aged 13 years was seen at the Medical College Hospital, Kottayam, with swelling over the sclera, proptosis and impairment of vision in the left eye of 3 months' duration.

Examination.—Her general health was normal. The left eye was proptosed, and medial and lateral movements were limited (Fig. 1). There was a painless, pinkish, vascular, fleshy growth on the lateral side of the orbit involving the lateral third of the lower fornix, the lateral canthus, and the lateral half of the upper fornix. Anteriorly it extended about 4 mm. from the lateral limbus. The swelling was covered by the conjunctiva and appeared to be circumscribed. The visual acuity was 6/60 and the fundus showed slight papilloedema with venous engorgement.

FIG. 1.—Part of growth on lateral aspect of orbit.

Operation.—A clinical diagnosis of angioma was made and the tumour was excised. The circumscribed partially-encapsulated tumour was found to have infiltrated the lateral rectus muscle. Profuse bleeding made it necessary to remove the posterior part of the tumour piecemeal. Orbital exenteration was performed and the tumour was seen to involve the whole orbit and to surround the optic nerve. A few months later the tumour recurred.

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Pathology.—Irregular pieces of pinkish fleshy tissue were available for histological examination.

A series of sections showed a uniform microscopic appearance. The tumour was composed of large polygonal cells with eosinophilic granular cytoplasm and spherical nuclei. In some areas the cells showed vacuolated cytoplasm (Fig. 2). The pattern was one of solid alveoli of cells surrounded by a thin vascular fibrous stroma (Fig. 3). A tubular arrangement of one or two rows of cells was seen in places. The nuclei of some cells presented a bizarre appearance. There were areas of necrosis. Sections of the lateral rectus muscle showed tumour invasion.

FIG. 2.—Photomicrograph, showing large polygonal cells and thin vascular fibrous stroma. Haematoxylin and eosin. $\times 450$.

FIG. 3.—Photomicrograph, showing characteristic pattern (reticulum). $\times 400$.

Discussion

Malignant non-chromaffin orbital paragangliomata produce alarming symptoms of proptosis and limitation of eye movements, but are extremely rare (Table). However, the literature contains reports of other orbital tumours interpreted as malignant granular

TABLE
INCIDENCE OF ORBITAL MALIGNANT NON-CHROMAFFIN PARAGANGLIOMA

Authors	Date	No. of Cases		
		Orbital	Other Sites	Total
Murphy and others	1949	0	175	175
Shuman	1961	1	49	50
Vakil and Sirsat	1963	1	10	11
Indira and others	1965	0	2	2
Bhargava and others	1965	0	2	2
Total		2	238	240

cell myoblastomata (Hogan and Zimmerman, 1962), non-chromaffin paragangliomata (Fisher and Hazard, 1952; Lattes, McDonald, and Sproul, 1954; Tye, 1961; Nirankari, Greer, and Chaddah, 1963), and even adrenal-rest tumours (Hughes and Ambrose, 1944) which were possibly all non-chromaffin paragangliomata.

There is no uniformity of opinion regarding nomenclature and histogenesis. Stout, (1953), Zimmerman, Netsky, and Davidoff (1956), Smetana and Scott (1959), and Willis

(1959) believed the tumour to arise from microscopic chemoreceptors similar to the paragangliomata arising from the carotid and aortic bodies. This was contradicted by Marshall and Horn (1961), who studied the morphological differences between non-chromaffin paragangliomata and alveolar soft tissue sarcomata. Christopherson and others (1952) considered them to be malignant soft tissue tumours of undetermined origin and described them as "alveolar soft-part sarcomata". The tumour has also been reported as a malignant granular cell myoblastoma. Ross, Miller, and Foote (1952) proved that the granular cell myoblastoma was different from the alveolar soft-part sarcoma. Moreover, the electron microscopic studies of Fisher and Hazard (1952) and Shipkey, Lieberman, Foote, and Stewart (1964) suggest that the granular cell myoblastoma arises from Schwann cells, whereas the histogenesis of the alveolar soft-part sarcoma is still unknown. The latter, however, is a distinct entity with the characteristic alveolar histological structure (made prominent by reticulum stains), and histochemical studies (Fisher, 1956; Hamperl and Lattes, 1957), P.A.S., and stains for acid mucopolysaccharide are often helpful. Intracytoplasmic crystalloid structures when seen are diagnostic.

The tumour should be differentiated from metastasis of renal carcinoma, hepatocellular carcinoma, and rare cases of orbital metastasis of pheochromocytoma. The tumour infiltrates locally, but distant metastases may occur later and may involve the lungs, brain, and bones.

Summary

A case of malignant non-chromaffin paraganglioma is reported, and the histogenesis and differential diagnosis are briefly discussed. This type of tumour should be remembered in cases of proptosis caused by orbital neoplasms.

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