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Chemodectomas

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SUMMARY

A pathological study of six chemodectomas seen during nine years period is presented. The histopathology, diagnosis, and histogenesis are discussed.

INTRODUCTION

A chemodectoma is a highly characteristic tumour which arises, most commonly, from the carotid body and glomus jugulare. Various terms have been used to describe them, e.g. nonchromaffin paraganglioma, endothelioma, granular cell myoblastoma, etc. Mulligan (1950) suggested the term 'chemodectoma' since the glomerae from which these tumours originate are chemoreceptors reacting to changes in blood PH.

Chemodectomas are rare, only 300 cases being described in the world literature. Six cases seen in nine years (1968 - 1976) in the Department of Morbid Anatomy, School of Medicine, Lusaka are presented.

CLINICAL DATA

Of the six patients two were men and four women. The men were aged 11 and 52 years, the women 20, 21, 25 and 28. They had presented with a slow-growing mass near the angle of the jaw, of maximum duration four years. The biopsy or the whole tumour was sent to the laboratory with various clinical diagnosis, e.g. salivary gland tumour, tuberculous, lymphadenopathy, carotid body tumour, metastasis, branchial cleft cyst, fibroma, etc.

PATHOLOGIC FINDINGS

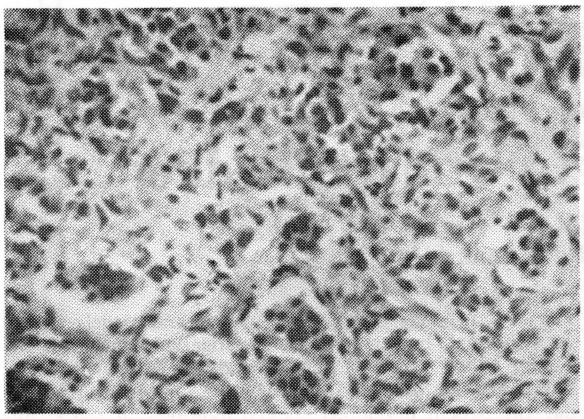
The tissue received were gray, pinkish, brown or reddish-brown. They were either fragmentary or lobulated. They had rubbery or firm consistency. They measured from 2.8 x 2.2 x 2 cm to 4.5 x 3.5 x 2 cms.

Microscopically, glomus jugulare tumours were identical to carotid body tumours. All the above biopsies showed nests of rather large polyhedral cells (Fig. 1). Reticulin fibres were not seen about individual cells within the nests (Fig. 2). Tumour cells were also arranged in columns and strands with a close relation to dense or scanty fibrovascular stroma. These cells were round or spindle-shaped with hyperchromatic nuclei. This pattern was observed predominantly in two cases. Groups of tumour cells were also arranged around vascular spaces in a peritheliomatous pattern in three cases. Rarely vascular spaces were large and looked like cavernous

hemangioma.

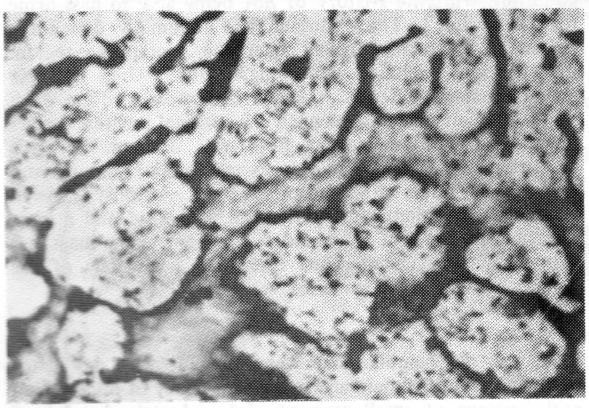
Cellular pleomorphism was observed in three tumours (Fig. 3). Lumina or walls of blood vessels or lymphatics were clear of tumour cells. There was a fibrous capsule in some cases. Ganglion cells were absent, but nerve fibres were seen at the periphery of tumour and in the fibrous septa. Fibrous trabeculae, some large and densely hyaline, and of varying sizes were seen dividing the tumours into irregular lobules in three cases, while foci of calcification were seen in one. Overall, the above histological appearances confirmed the diagnosis of chemodectomas in all six cases.

FIG. I



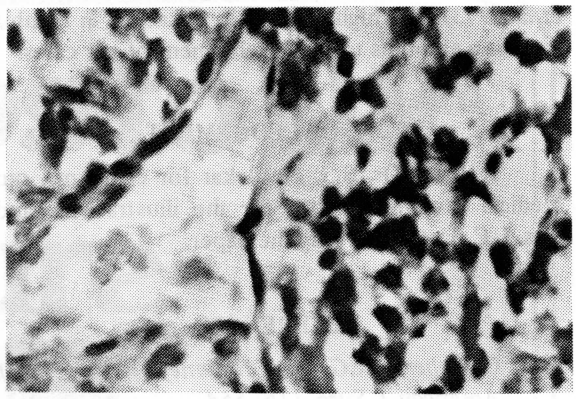
'Organoid' appearance of chemodectoma formed by nest of tumour cells surrounded by vascular connective tissue stroma. The nuclei are oval with distinct nucleoli; the cytoplasm is pale, eosinophilic and vacuolated; cell membranes are distinct (H and E x 444)

FIG. II



Reticulin stain of tumour from cases four to emphasize 'organoid' pattern and no reticulin in-between tumour cells in nests.

FIG. III



Cellular pleormorphism and relationship of tumour cells to an endothelial-lined blood channel.

Pleomorphism is moderate; mitotic figures are rare. (H and E, X 888)

DISCUSSION

Chemodectomas may develop at any age, but the commonest age-incidence is 30 to 40 (Evans, 1966). In contrast, five of our six cases were young (11 - 28). Chemodectomas occurs with equal frequency in both sexes (Evans, 1966). The present series show female preponderance. Chambers and Mahoney (1968) pointed out high (30%) preoperative misdiagnosis rate. However, the histological appearances are usually diagnostic (Evans, 1966), and cell clusters separated and isolated like islands by the well vascularized fibrous septa, rarity of mitotic figures, and reticulin stain, being characteristic. The histopathological study lacks prognostic value, because, whereas tumours with a regular cytologic pattern may metastasize, those showing marked cellular pleomorphism may remain benign (Romanski, 1954).

These tumours grow slowly, sometimes 10 to 20 years. Metastases are rare, but may occur in the regional lymphnodes, bone, liver, lung and heart. Glomus jugulare grows slowly but is locally destructive. It may recur following excision but metastases are rare. Occasionally, multicentric tumours are reported in patients with a familial history (Guild 1953).

They are neurectodermal in origin, being distinct from the endothelial lining of capillaries or sinusoids. Pryse-Davies *et al* (1964) concluded that the chief cell, and ganglionic and paraganglionic elements found in normal carotid bodies, could be variously reproduced in chemodectomas. One observa-

tion suggests that carotid body tumours may be more frequent in people living at high altitude (Saldna *et al* 1973). The high attitude hypoxaemia in Zambia may be a stimulus to the development of our cases.

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