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Melanotic spinothoracic schwannoma

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ABSTRACT A dumb-bell mediastinal melanotic schwannoma is described, and the rarity of this type of tumour emphasised. The tumour was resected by simultaneous laminectomy and posterior thoracotomy. The patient is well four years after operation. The pathology of the tumour is described and the origin of melanotic cells is discussed.

Melanotic schwannoma are rare nerve sheath tumours. Since the first description of this neoplasm by Björneboe (1934) in a patient suffering from Von Recklinghausen's disease, only 20 cases have been reported (Mandybur, 1974). The tumours have usually been found in the subcutaneous tissue.

Although primary melanotic tumours have been described in the thorax, especially in the oesophagus where Piccone *et al* (1970) referred to 30 examples, they are occasionally described in the lung (Taboada *et al*, 1972), in the heart (Gelfand *et al*, 1977), or in the mediastinum. We have found only six papers describing melanotic pigmented tumours of the mediastinum. In four cases the pigmented tumour arose from ganglion cells (Millar, 1932; Kellert and Woodruff, 1956; Gautam, 1974; Hahn *et al*, 1976) and two were spinothoracic melanotic schwannoma (Mandybur, 1974; Bagchi *et al*, 1975).

We describe a patient who had a dumb-bell mediastinal melanotic schwannoma extending from the right upper mediastinum into the spinal canal through the intervertebral foramen.

Case report

A 49-year-old woman was admitted to hospital suffering pain in the right arm in the distribution of the eighth cervical nerve root. This had started two years previously, but for the past six months the pain had been severe and continuous.

Neurological examination did not show any motor, trophic, or sensory changes in the limbs. A chest radiograph showed a mass in the right upper mediastinum (fig 1) suggesting a neurological tumour. Spinal films showed widening of the intervertebral foramen between the seventh cervical and first thoracic vertebrae; and myelography (fig 2) showed a lateral deformity of the contrast column on the right side. Vertebral arteriography showed a displacement of the first portion of the

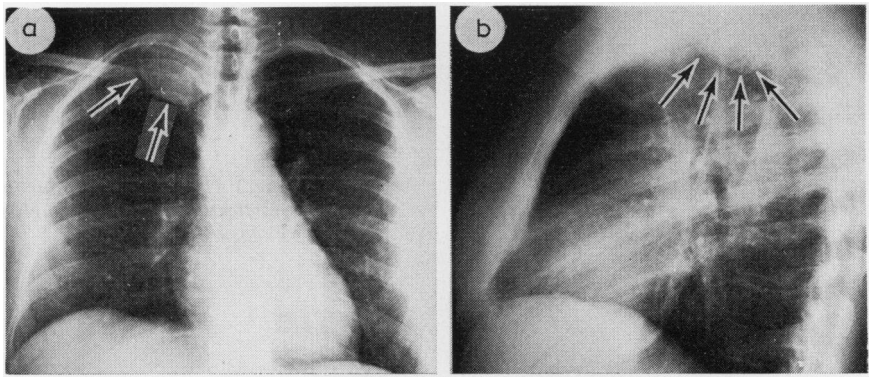

Fig 1 *Anteroposterior (a) and lateral (b) chest radiographs showing a right-sided neurogenic mediastinal tumour.*

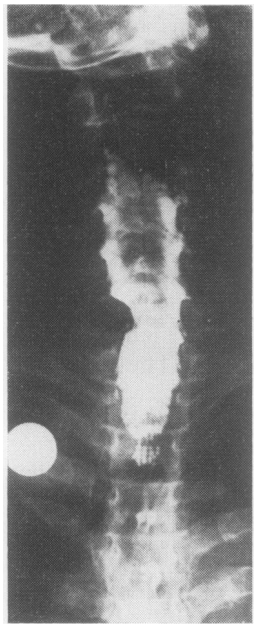

Fig 2 *Myelogram. Deformity of contrast column on right side delineates intraspinal portion of tumour.*

right vertebral artery.

In May 1974 a laminectomy (sixth cervical to first thoracic) was performed simultaneously with a posterior thoracotomy with the patient in the prone position on the operating table. A well-encapsulated black pigmented extradural tumour was found. It arose from the eighth cervical nerve root and extended through the intervertebral foramen towards the mediastinum, where the principal tumour mass lay (fig 3). The nerve root was sectioned, and the tumour was completely and easily removed through the combined exposure. The postoperative recovery was uneventful, and the patient was discharged 15 days after operation.

Pathological examination showed a smooth encapsulated black tumour measuring 6·0×4·5×2·2 cm. The entire specimen was fixed in 10% formalin. A portion was embedded in paraffin and stained by standard techniques (haematoxylin and eosin, Masson trichromic, and Gomori reticulin). Prussian blue and Fontana's methods were used to identify pigmented granules. The DOPA reaction could not be tested because the tumour had been fixed in formalin.

On microscopy (fig 4), the tumour was shown to be circumscribed by a fibrous capsule. The central portion of the tumour was cellular showing plump spindle cells arranged in fascicles creating a pattern of whorls and palisades. The nuclei showed neither pleomorphism nor mitotic activity. Many cells were filled with variable amounts of small black pigmented granules making nucleus identification difficult. These granules were identified as melanin (Fontana stain positive, iron negative). The stroma was scarce and showed fibrous bundles and similar pigmented granules.

Secondary phenomena seen were blood vessels with thickened walls, extensive haemorrhagic areas, and macrophages containing dark yellow iron positive granules. Neither calcium deposits nor microcysts were present.

Despite the adequacy of surgical resection the patient was treated by irradiation (5500 rads) over an eight-week period. Four years later she showed no clinical or radiological signs of recurrence of the tumour.

Discussion

Melanotic schwannoma is a rare neoplasm with a

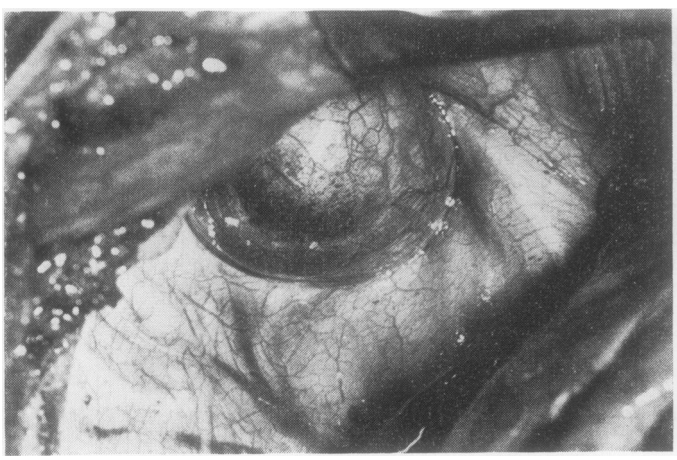

Fig 3 *Pigmented tumour in posterior mediastinum seen through transthoracic exposure.*

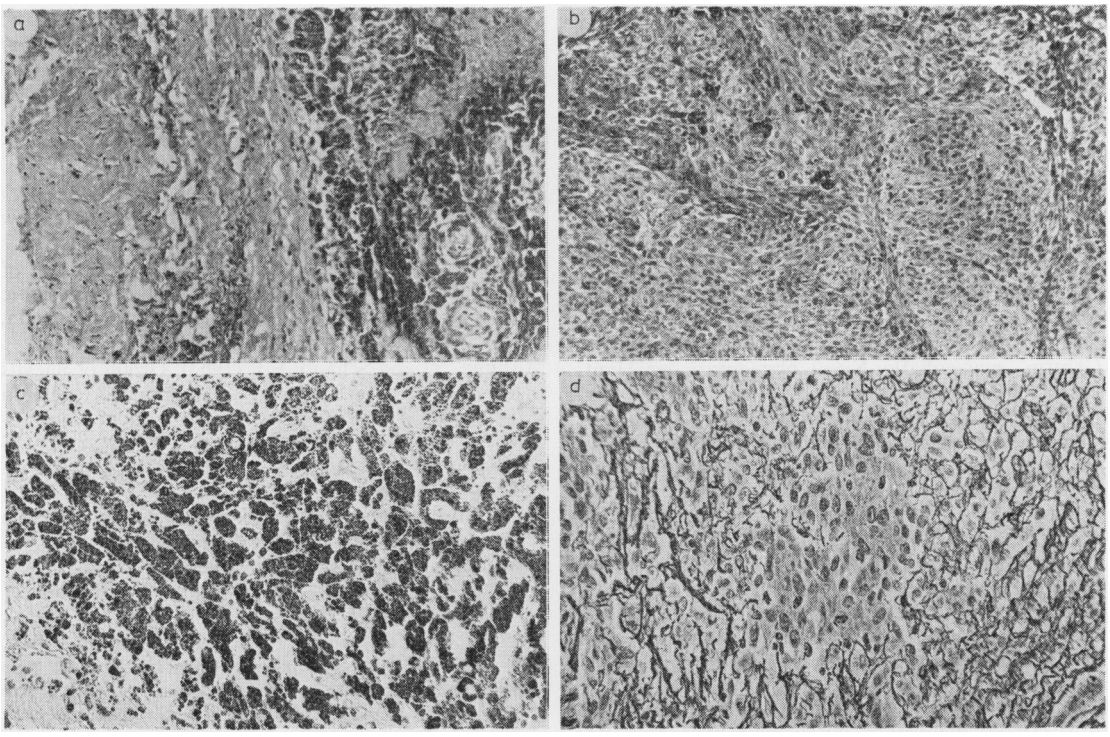

Fig 4 *Tumour histology: (a) On left the nerve sheath which constitutes capsule of tumour (Haematoxylin and eosin ×58); (b) plexiform pattern of tumour (H and E ×92); (c) tumour cells with melanotic intracytoplasmic granules (Fontana ×144); and (d) network of reticulin fibres surrounding cells (Gomori reticulin ×144).*

controversial histogenesis because of the uncertain origin of the melanin pigment. In our case microscopy showed two cell types, one non-pigmented, having the cytological characteristics and architectural features of a schwannoma, the other showing melanin granules.

Hodson (1961) stated that the presence of ectopic melanotic cells was the result of a failure of complete migration of these cells; thus they were not reaching their cutaneous destination during embryologic development. Mandybur (1974) has shown by electron microscopy the presence of cells with melanosomes in an intraspinal tumour. He explained the histogenesis of melanotic nerve sheath tumours as originating from aberrant or ectopic melanotic cells.

Dastur *et al* (1967) suggested that the presence of melanin in schwann cells was caused by the phagocytosis of pigment elaborated by melanoblasts. Schwann cells have been found to have phagocytic activity in tissue cultures (Lumsden, 1963).

The melanocytes when present in the nerve sheath can transfer their melanin to the schwann cell. Masson (1948) considered the melanocyte as a glandular cell producing melanin and suggested the term "cytocrine activity" to describe the passage of insoluble melanin granules directly to the cytoplasm of epithelial cells. Hodson (1961) states that the melanocyte embraces the epithelial cell and deposits its melanin by means of secondary branches extending from the dendrites to enter the receptor cell.

Another hypothesis to explain the histogenesis of this tumour is based on the possibility of melanomatous transformation of the schwann cell. The nerve sheath cell and the melanocyte have a similar histological origin from the neural crest cell (Mishima and Schaub, 1963; Bird and Willis, 1969). Nakai and Rappaport (1963) induced melanotic tumours in the hamster and found neoplastic schwann cells in these tumours. This supports the theory of the transformation of the schwann cells into melanocytes. Shillitoe (1965), unable to detect the presence of melanocytes in melanotic schwannoma, concluded that the schwann cell alone could produce melanin. This concept is supported by Hahn *et al* (1976) who

showed by electron microscopy the existence of melanosomes in different stages of development in the schwann cells of a pigmented ganglioneuroblastoma.

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