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Multiple ganglioneuroma of the neck

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Abstract
A case of multiple ganglioneuroma arising along the entire length of the cervical sympathetic chain of one side of the neck is described. This is a distinctly unusual site and distribution of the disease and computed tomography proved invaluable to demonstrate its extent, in addition to excluding involvement of more caudal regions.

Case report

A 6.3-year-old child presented with a four month history of a gradually enlarging swelling of the left neck. There were no infective upper respiratory tract or systemic symptoms and no relevant family history. Physical examination demonstrated a clinically solitary firm mobile 4 × 3 cm tumour located in left mid-neck presenting beneath the anterior border of sternomastoid. This was thought to be a lymph node. General clinical examination was normal. A preliminary biopsy revealed a ganglioneuroma which microscopically appeared as a tumour composed of clusters of ganglion cells contained within a background of Schwann cell bundles (Fig. 1), confirmed as such by immunocytochemical staining for S100 neuro-protein. Whole body computed tomography (CT) suggested that the disease was confined to the right side of the neck and comprised a chain of multiple well encapsulated tumours which extended from the skull base to the root of the neck. The lower extent of disease is shown in Figure 2. This was confirmed on surgical exploration where six discrete masses were removed (Fig. 3). A post-operative Horner's syndrome was noted.

Comment

Ganglioneuroma is an uncommon tumour of the sympathetic

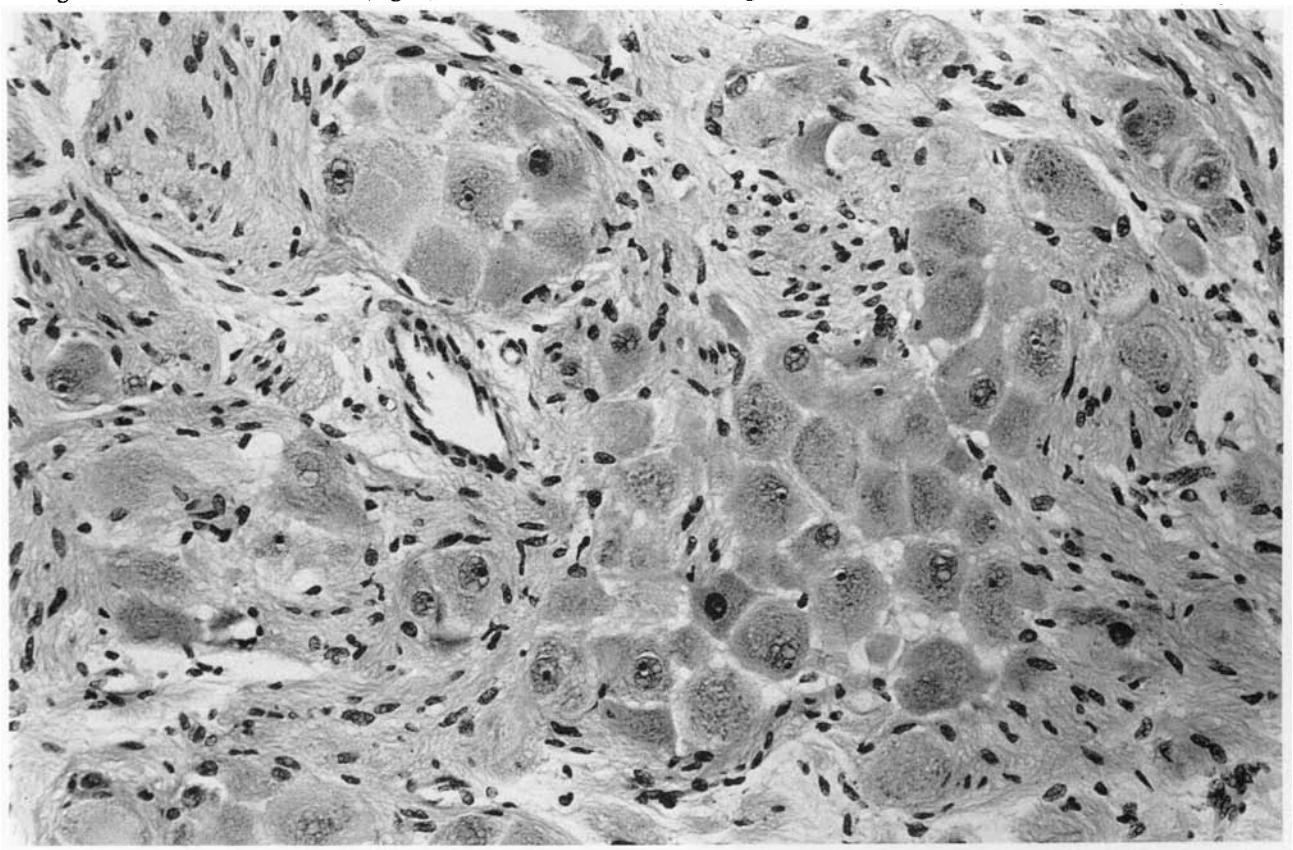

FIG. 1
Photomicrograph of histopathological section, high power, ×400 magnification.

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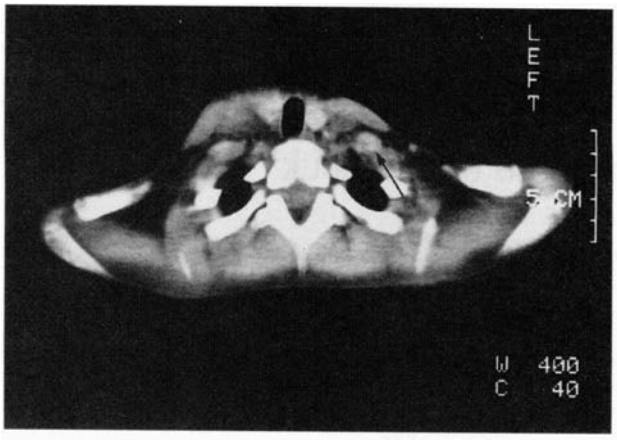

FIG. 2

Computed tomogram of lower neck demonstrating the lower extent of the left neck disease, arrowed.

FIG. 3

Macroscopic view of excised tumours.

nervous system and generally presents as a solitary mass, usually in the mediastinum or abdomen (Stout, 1947; Enzinger and Wies, 1986). They are less frequently encountered in the neck and multiplicity at this site is very rare. Despite this, certain recognized associations exist which make further evaluation beyond simple excision mandatory: spontaneous cytomaturation of metastatic neuroblastoma into ganglioneuroma has been described (Garvin *et al.*, 1984; Ricci *et al.*, 1984) and therefore the whole of the sympathetic chain and adrenal glands should be visualized to exclude tumours at other sites. CT is clearly the tool of choice (Armstrong *et al.*, 1982); malignant change within a ganglioneuroma has been documented (Garvin *et al.*, 1984; Ricci *et al.*, 1984) and therefore complete excision of all tumours should be undertaken. Finally, the vasoactive properties of ganglioneuroma tissue are also recognized (Trump *et al.*, 1977; Mendelsohn *et al.*, 1979), although watery diarrhoea, and symptoms attributable to hypokalaemia and hypochlorhydria would be anticipated in these cases.

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