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Cervical ganglioneuroblastoma

by

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Ganglioneuroblastoma is a differentiated form of neuroblastoma. This malignant embryonic tumour of neural crest origin can involve the sympathetic ganglia throughout the body. Primary neuroblastoma of the cervical ganglion is rare; its differentiated form, ganglioneuroblastoma, is even more rare. Only one previous case to our knowledge has been documented.

Case Report

C.D. was referred to the ENT Department at the age of 2½ years with a history of recurrent tonsillitis and a left cervical mass which had appeared with the first attack of tonsillitis six months earlier; there were no other symptoms or signs. Clinical examination revealed enlarged unhealthy looking tonsils and a swelling in the left side of the neck anterior to the sternocleidomastoid muscle, thought to be lymphadenitis, secondary to tonsillitis. Full blood count and ESR were normal. Tonsillectomy was not recommended because of his age. Two years later the child was referred again because of increase in the frequency of tonsillitis and the enlarging cervical mass, but again no other symptoms or signs, and full blood count and ESR were normal. At this time it was decided to perform adeno-tonsillectomy, and while the child was anaesthetised and fully relaxed, the swelling was noted to be soft and was displacing and bulging into the pharynx medially. The clinical diagnosis was thought to be a branchial cyst, and so adenotonsillectomy was not carried out. A week later the neck was explored and a fairly large, soft and clinically cystic lesion was found in the carotid sheath pushing the carotid vessels (common, internal and external) and the vagus medially and extending deep to the vagus but superior to the transverse process of the atlas; there were a few lymph nodes lying adjacent to the mass. The mass together with the lymph nodes were dissected completely from the surrounding structures without difficulty.

Macroscopically the mass was firm, solid and encapsulated (Fig. 1) measuring 5.5 × 4.0 × 3.0 cm. and weighing 35 gms. Histological examination revealed that this was an encapsulated ganglioneuroblastoma. The tumour was composed predominantly of immature neuroblasts with small rounded cells and hyperchromatic nuclei (Fig. 2). In areas there was evidence of maturation with a fibrillary matrix resembling glial tissue containing larger ganglion cells (Fig. 3) in some of which Nissl granules were clearly seen (Fig. 4). The fibrous capsule was intact and within it a sympathetic ganglion was identified. The lymph nodes lying on the mass showed reactive changes and no evidence of tumour.

Post-operatively the child developed a classical ipsilateral Horner's syndrome but otherwise his recovery was uneventful. Further investigations failed to reveal evidence of either an occult primary site or disseminated disease. Abdominal and chest X-rays, repeat full blood count, bone marrow, abdominal ultrasound, bone scan, skeletal survey and 24-hour urinary VMA were all normal. In view of the localized nature of the tumour and its complete excision, no further treatment has been given. He remained well and free of disease 3 years after diagnosis.

Discussion

Neuroblastoma is a malignant tumour of neural crest origin and is a member of the family of neoplasms of the APUD neuroendocrine system (Bolande, 1974). It is probably the most frequent solid malignant tumour in infants and young children, 46 per cent occurring below the age of two years and 83 per cent below the age of seven years (de Lorimer *et al.*, 1969), and although it most

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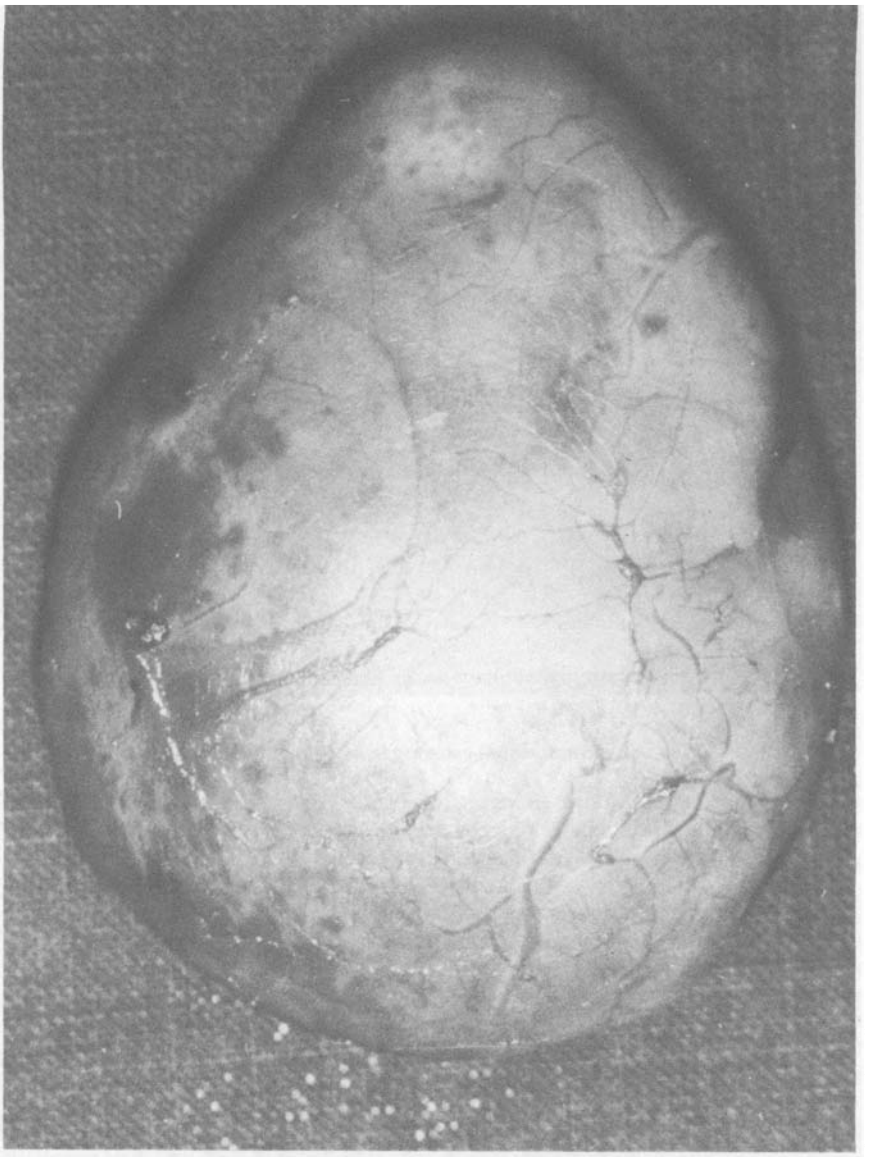


FIG. 1

Excised tumour with a smooth capsular surface.

commonly originates in the adrenal medulla it may arise in any of the sympathetic nervous system ganglia. Between 75–90 per cent of these tumours arise in the abdomen and 10–15 per cent within the thoracic cavity (Brown *et al.*, 1978). Although the occurrence of metastases in the neck is well established (Brown *et al.*, 1978), a primary tumour in the neck is rare (Conley, 1955; Brown *et al.*, 1978). Table I lists the cases of primary cervical neuroblastoma recorded in the literature since 1959 together with the total number of cases analysed from all sites. Including the present case, the incidence of a primary tumour in the neck is less than 5 per cent.

The presence of a mass, high or low, in the neck without any evidence of its neurogenic origin is the most frequent presenting complaint. Signs and symptoms of compression of adjacent structures

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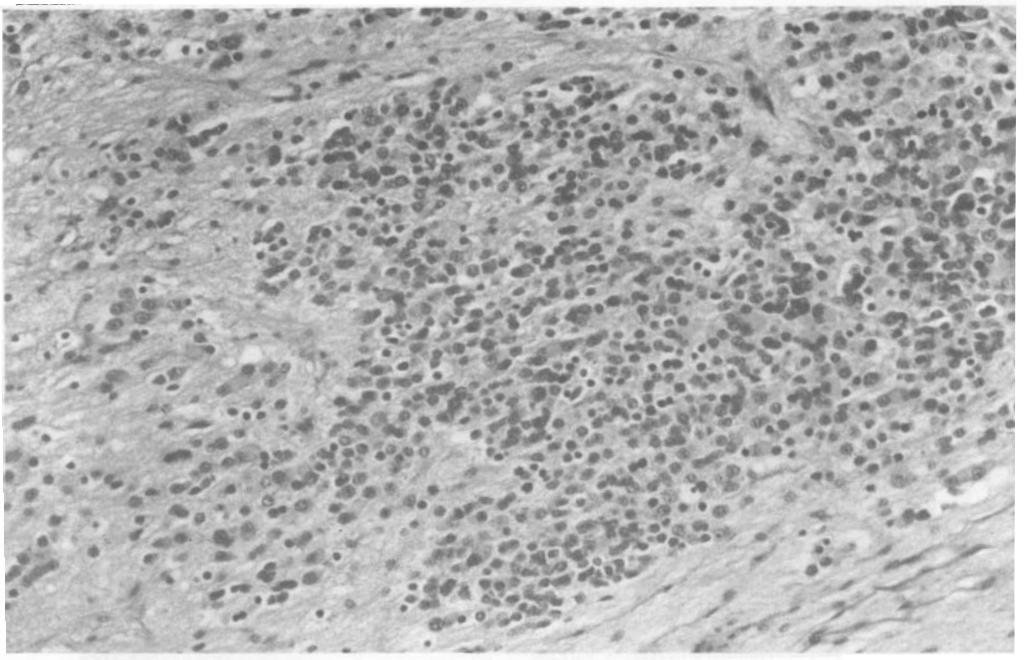


FIG. 2

Immature neuroblasts in a fibrillary matrix (haematoxylin and eosin, ×100).

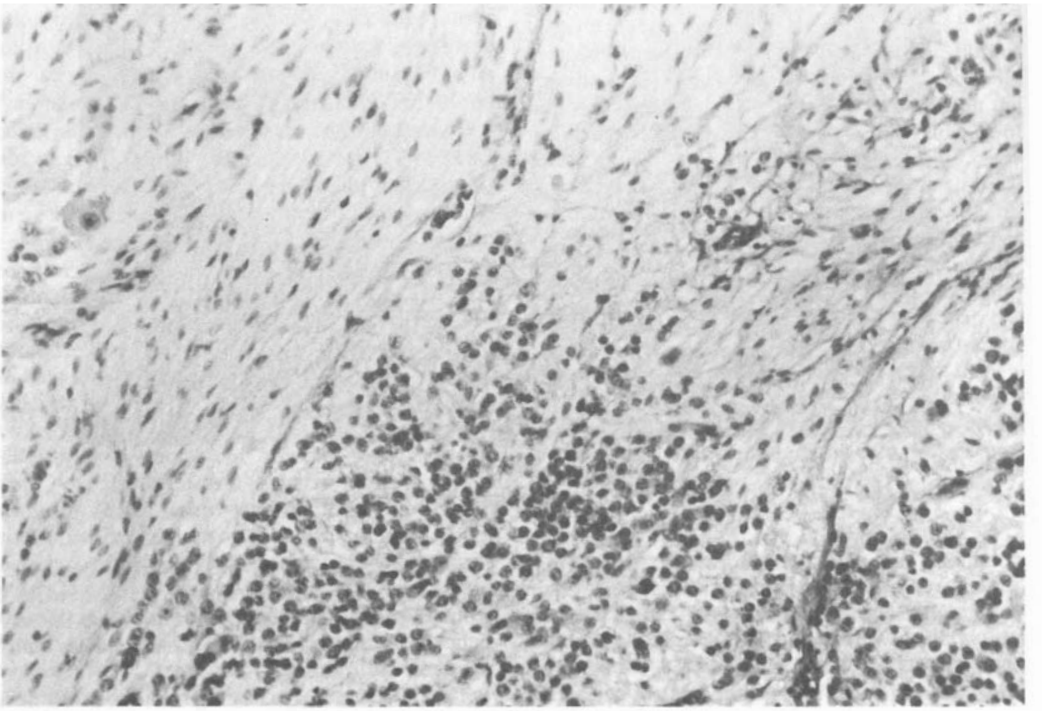


FIG. 3

Fibrillary matrix containing a ganglion cell (haematoxylin and eosin ×100).

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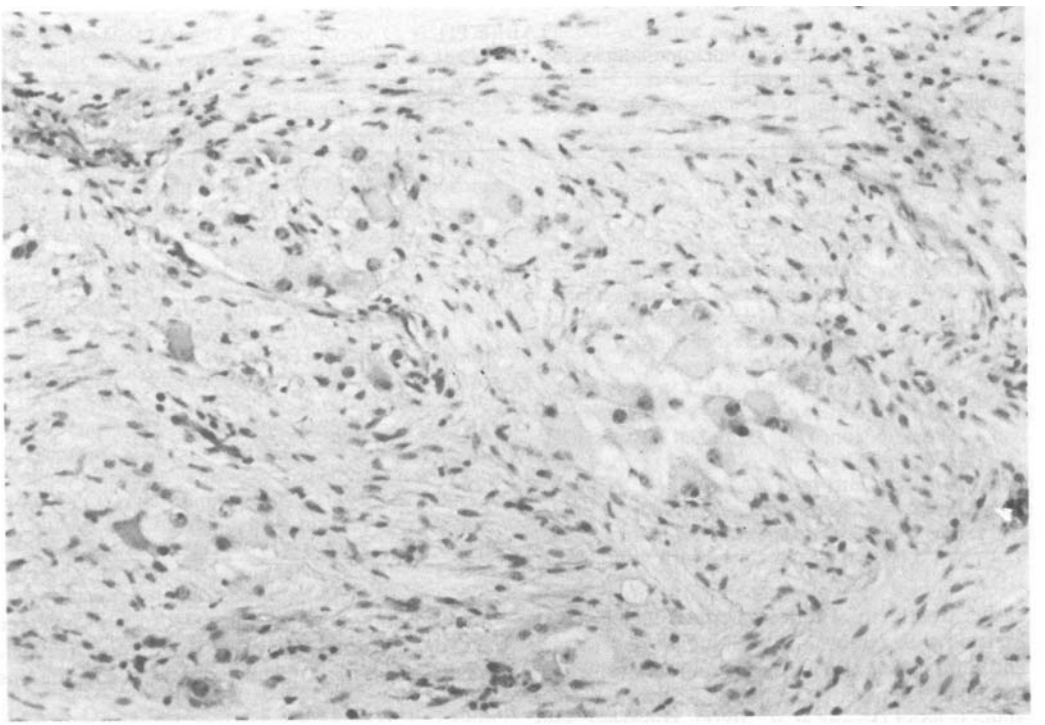


FIG. 4

Ganglion cells containing Nissl granules (haematoxylin and eosin ×100).

such as the sympathetic nerve chain, oesophagus, larynx or trachea have been present in most recorded cases, but were absent in this case. Ipsilateral Horner's syndrome and heterochromia of the irides has been documented and both may develop before the discovery of the tumour (Jaffe *et al.*, 1975). Weakness in the upper limb due to compression of the cervical and upper thoracic nerves has been reported (Gilboa and Campbell, 1976) and also, less frequently, dumbbell extension into the vertebral canal (King *et al.*, 1975).

In view of the fact that neuroblastoma may metastasise to the neck and may even be the presenting symptom (Brown *et al.*, 1978), the decision that a cervical neuroblastoma is primary rather than secondary can only be made after exclusion of a primary site elsewhere in the body. Important investigations must include abdominal and chest X-rays, intravenous pyelography and bone marrow examination. The development of computerised tomography has now been shown to be of great value in diagnosis (Armstrong *et al.*, 1982) and in this case confirmed the absence of tumour in any other site.

Neuroblastoma is a tumour in which the prognosis must be guarded. Factors which influence the prognosis include the age of the child (Kinnier Wilson and Draper, 1974), the extent of the tumour, the histopathological type and its site and treatment.

The vast majority of neuroblastomas, whatever the site, are undifferentiated in type. The presence of areas of differentiation into ganglion cells, ganglioneuroblastoma, has been regarded as evidence of maturation of the tumour and associated with a better prognosis (Kinnier Wilson and Draper, 1974). Whether this is so probably depends on the degree of differentiation of the neuroblastoma element of the ganglioneuroblastoma (Fasicle). The tumour in this case was composed predominantly of primitive cells, undifferentiated neuroblastoma tissue, and it is relevant to note that the only other recorded case of primary cervical ganglioneuroblastoma (Rosenfeld *et al.*, 1968) died one year after the onset of symptoms.

Regardless of the degree of histopathological differentiation, the evidence available in the literature does indicate that the prognosis of primary cervical neuroblastoma is better than expected in neuroblastoma arising in other sites (de Lorimer *et al.*, 1969; Orr, 1978). This may be explained by the likelihood of earlier diagnosis and consequent

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TABLE I
CERVICAL GANGLIONEUROBLASTOMA INCIDENCE OF CERVICAL NEUROBLASTOMA

Year	Author	No. of cervical cases	Neuroblastoma cases reported
1959	Gross *et al.*	5	217
1959	Bodian	7	129
1962	D'Argeon	4	236
1964	Sutow	1	3
1967	Priebe and Clatworthy	2	92
1968	Rosenfeld *et al.*	1	1
1968	Fortner *et al.*	3	133
1969	de Lorimer *et al.*	27	212
1970	Young *et al.*	3	35
1971	Swank *et al.*	5	125
1973	Jaffe and Jaffe	3	178
1976	Jones and Campbell	1	144
1978	Orr	3	3
1978	Brown *et al.*	7	105
1982	Cushing *et al.*	6	6
1984	Al-Jassim	1	1

earlier treatment. The treatment of choice is wide excision with removal of local lymph nodes (Jaffe and Jaffe, 1973) and in Stage I tumours, which are completely excised, the prognosis with surgery alone is good (Evans *et al.*, 1971). However, post-operative radiation is generally used (Stella *et al.*, 1970) and, in more advanced cases, is combined with chemotherapy (Evans *et al.*, 1976). Elevated levels of urinary catecholamines, which are known to be important in both the diagnosis and response to treatment of neuroblastoma, have been reported as normal in the primary cervical tumours (Evans *et al.*, 1976; Orr, 1978). This probably reflects the lack of investigation prior to the removal of the tumour, as in this case, and such investigation should be part of the long-term follow-up as an early indicator of recurrent disease.

Summary

A case of primary cervical ganglioneuroblastoma is reported. It is a very rare tumour, only one example of this differentiated form of neuroblastoma having been previously recorded as a primary tumour in the neck. The incidence, clinical features, pathology and prognosis of primary cervical neuroblastoma have been discussed.

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