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Primary cervical ganglioneuroblastoma

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ABSTRACT

Objectives: Ganglioneuroblastomas represent a histological subgroup of the rare neuroblastic tumours with intermediate malignant potential arising from neural crest progenitor cells of sympathetic nerves. Diagnosis can often be difficult based on imaging alone. We describe 4 cases of children presenting with a solitary neck mass with histology ultimately revealing ganglioneuroblastoma.
Methods: A retrospective case note review was carried out of all patients with cervical ganglioneuroblastoma seen at Great Ormond Street Hospital, UK.
Results: Mean age at presentation was 5 years. Based on imaging, the initial diagnoses for three of the cases were: lymphatic malformation, carotid body tumour, paraganglioma, respectively, whilst the remaining case had an immediate incisional biopsy revealing the correct diagnosis. All cases were managed by surgical excision with no evidence of recurrence after a median follow up of 6 years.
Conclusion: Otolaryngologists should be aware of ganglioneuroblastoma when establishing the differential diagnosis of a child presenting with a neck mass. Biopsy is recommended as the gold standard investigation to avoid an incorrect diagnosis.

1. Introduction

The vast majority of solitary neck lumps in children are cervical lymph nodes which generally resolve without intervention and, unlike adults, need no further cytological investigation. Imaging can be very useful in identifying the very small subset of children whose neck lump represents a neoplasm, yet the radiological diagnosis may not always be certain.

Between 2009 and 2014 our unit treated 4 children with cervical ganglioneuroblastoma. These tumours represent a histological subgroup of the rare neuroblastic tumours which arise from neural crest progenitor cells of sympathetic nerves. They are composed of both mature gangliocytes and immature neuroblasts and have intermediate malignant potential, with less malignant potential than neuroblastoma but more so than ganglioneuroma. In the neck presentation is usually with a lateral neck mass or retropharyngeal mass though patients may present with Horner's syndrome.

Over the last few decades, very few cases of paediatric cervical ganglioneuroblastoma have been reported [1,2]. An epidemiological study in the USA estimated the incidence of neuroblastic tumours as a whole to be 7.6 per 1 million [3]. Cervical involvement is thought to account for 5% of all neuroblastic tumours [4], therefore within this group ganglioneuroblastomas originating in the neck are exceptionally rare. Despite advances in imaging techniques we show in our series that the diagnosis can initially be incorrect. We discuss our management and outcomes and compare this to previous literature.

2. Case reports

2.1. Case 1

A 4 year old female child presented with an enlarging right neck mass, cough and fever. Ultrasound scan revealed a well-defined and well circumscribed mainly homogenous mass within the right neck extending from the level of the angle of the mandible inferiorly, measuring approximately 5.8 cm × 4.1 cm × 3.1 cm. The same lesion was seen on MRI scan displacing the carotid artery anteriorly (Fig. 1). Incisional biopsy was performed with histology revealing intermixed schwannian stroma rich ganglioneuroblastoma. There was no lymph node involvement or N-myc amplification on FISH study. Laboratory values for full blood count, electrolytes, coagulation, liver function, magnesium and ferritin were all within normal range. Urinary catecholamines were

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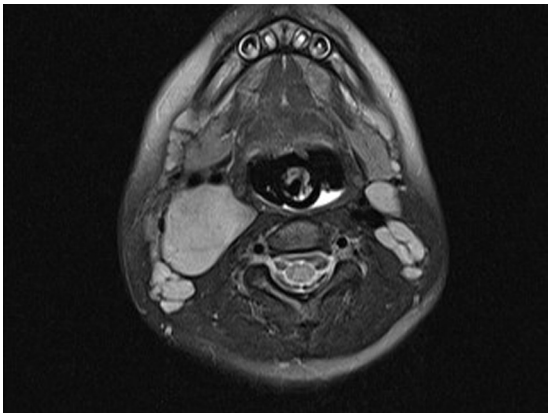

Fig. 1. Case 1 MRI showing 5.5 cm × 3.8 cm × 3.1 cm mass lesion in the right neck displacing the carotid artery anteriorly. The right internal jugular vein is effaced.

within normal range. Bilateral bone marrow aspirates and trephines showed no evidence of metastatic infiltrate. 123I-MIBG scan showed no selective uptake of MIBG.

A multidisciplinary decision was taken to observe without intervention initially. At 3 months ultrasound revealed a small reduction in tumour size but at 6 months another ultrasound showed an increased size. Surgical excision of the tumour was undertaken. The mass arose from cervical sympathetic nerves and lay deep to the right carotid artery in the parapharyngeal space. Accessory and hypoglossal nerves were identified and preserved. Post-operatively the patient had a right Horner's syndrome.

Follow-up was undertaken at 6-monthly intervals for the first 3 years and thereafter annually with urinary catecholamines and ultrasound of the neck and abdomen. MRI scan at 18 months showed no sign of disease recurrence. At 5 years the patient was well with no sign of recurrence. The signs of the Horner's syndrome were barely noticeable.

2.2. Case 2

A 5-year-old male child presented with an enlarged left tonsil. Tonsillectomy was performed where a normal tonsil was found to be overlying a parapharyngeal mass. MRI revealed a cystic lesion in the left parapharyngeal space extending from the skull base to level III (Fig. 2). A diagnosis of lymphatic malformation was made and injection sclerotherapy was planned. During the sclerotherapy procedure, the mass was noted to be echogenic, heterogenous and solid. The procedure was abandoned and instead a biopsy taken, with histology revealing intermixed schwannian stroma rich ganglioneuroblastoma. There was no lymph node involvement or N-myc amplification on FISH study. Blood tests as per the previous case were all within normal range. Urinary catecholamines were within normal range. Bilateral bone marrow aspirates and trephines showed no evidence of metastatic infiltrate. 123I-MIBG scan showed no selective uptake of MIBG.

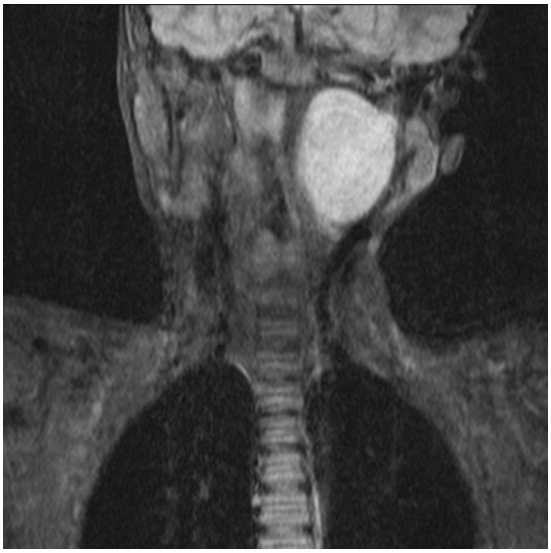

Fig. 2. Case 2 MRI showing Solid mass in the left parapharyngeal space displacing the left carotid sheath posteriorly and laterally and the pharynx medially.

Surgical excision of the tumour was undertaken via a cervical approach extended to a parotidectomy incision (Fig. 3a and b). The tumour arose from a thickened cervical sympathetic chain. Hypoglossal, facial and vagus nerves were identified and preserved. The accessory nerve was not disturbed. Tumour was separated from the internal jugular vein and internal carotid artery. Post-operatively the patient was found to have a facial palsy which subsequently recovered completely. Left Horner's syndrome was persistent. The patient also had reduced ipsilateral palatal movement which responded almost completely to speech therapy. Annual follow-up with MRI scan was undertaken. At 5 years the patient remains well with no sign of disease recurrence.

2.3. Case 3

An 8-year-old female child presented with a 6 month history of enlarging submandibular mass. MRI scan showed a large suprahyoid mass in the left parapharyngeal space and left carotid space, displacing the external and internal carotid arteries anteriorly and posteriorly respectively (Fig. 4). A small enhancing

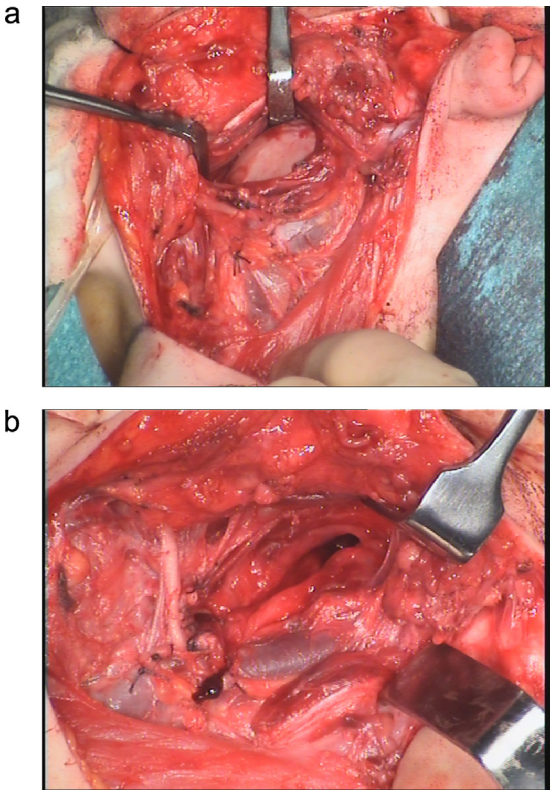

Fig. 3. (a) and (b) Case 2 excision of tumour via a cervical approach extended to a parotidectomy incision. The tumour arose from a thickened cervical sympathetic chain. Hypoglossal, facial and vagus nerves were identified and preserved. Tumour was separated from the internal jugular vein and internal carotid artery.