

Cervical Ganglioneuroma in a Child

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Abstract

Ganglioneuroma is a rare, benign, non-invasive and neurogenic tumor. These tumours usually present as a slowly enlarging mass, predominantly in single form. Due to its rarity and the lack of specific signs and symptoms, it is often difficult to reach a definite diagnosis prior to pathological examination. We are presenting a case of 18 month old child who presented with a neck swelling of short duration, which was completely excised.

Introduction

Ganglioneuroma are rare, benign tumours arising from the sympathetic system. They are usually seen in children and young adults less than 20 years of age with a slightly female preponderance [1]. Common sites are posterior mediastinum, retroperitoneum and adrenal glands and only 8% of the tumours occur in the neck. The usual presentation is an enlarging neck mass, but may occasionally present with signs of Horner's syndrome or due to compression of vital structures. We report a case of cervical ganglioneuroma arising in a 18 month old male child with a painless swelling in the neck of short duration. The mass was excised. The case is presented because of rarity of the condition at such a young age.

Case Report

An eighteen month old male child presented with a neck swelling of 2 months duration. The swelling was gradually increasing in size. There was no associated pain, fever or dysphagia. On local examination, a 2 cm x 3 cm swelling was found on left upper neck, which had smooth surface, was non-tender, mobile, and there was no movement on protrusion of the tongue (Figure 1a & 1b). Fine Needle Aspiration Cytology (FNAC) suggested a salivary neoplasm (Figure 2). The tumour was excised under general anaesthesia. Cut section of the tumour showed grayish white homogenous mass, no areas of haemorrhage or necrosis noted. It was well encapsulated. Histology showed well encapsulated benign tumour with groups of ganglion cells with large amount of cytoplasm, vesicular nuclei and prominent nucleoli. The background was fibrillary, composed of neural and fibroblastic tissues suggestive of ganglioneuroma (Figure 3). A small swelling in relation to the mass showed nerve trunks with ganglion cells.

Discussion

Ganglioneuroma was first reported by Loretz in 1870 and ganglioneuroma of the neck was first reported by De Quervain in 1899 [1]. Cervical ganglioneuroma are rare, benign tumors of neurogenic origin and accounting for about 6% of the childhood tumours [2]. The tumour commonly arises in the cervical sympathetic chain and often manifest with signs of Horner syndrome [3]. Ganglioneuroma is composed entirely of ganglion cells and Schwann and stroma [4]. 60% of these tumours arise under 20 years of age with slight female preponderance [1,4]. They are most commonly localized in the posterior mediastinum (41.5% of cases), retroperitoneum (37.5%), adrenal gland (21%), and neck (8%). Although they appear as asymptomatic mass in the majority of the cases, they may cause symptoms because of compression of vital structures in the neck and in about a third of the cases symptoms may appear because of high level of catecholamines [1,5]. Junli et al reviewed the English-language literature on ganglioneuroma over a ten year period and found only one case of multiple ganglioneuroma occurring on one side of the neck and bilateral neck masses were present in 3 cases [6].

Magnetic Resonance Imaging (MRI) and Computerized tomography are more valuable imaging techniques in the diagnosis of ganglioneuroma. MRI is superior to CT in the diagnosis of intraspinal tumors [7]. The tumor has been shown to have a capsule and calcifications both radiologically and histologically [5,8]. Although ganglioneuroma tends to be a more homogeneous tumor than neuroblastoma or ganglioneuroblastoma, it is not possible at imaging evaluation to discriminate among these three tumors [9].

Fine needle aspiration cytology (FNAC) is a rapid, cost-effective and safe diagnostic procedure, particularly in the pediatric age group [10,11]. However, results are often misleading, as was in the present case. Most characteristic histological feature of ganglioneuromas is the presence of



Figure 1a: Excised mass showing well encapsulated tumour with smooth surface.

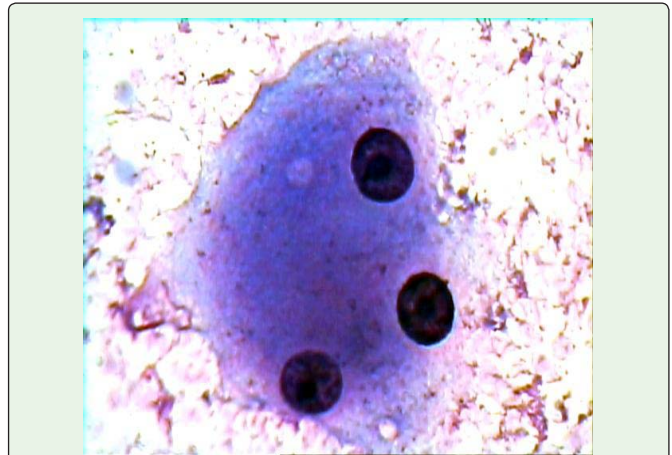


Figure 3: H&E stain 400x [High power]: well encapsulated benign tumour with groups of ganglion cells with large amount of cytoplasm, vesicular nuclei and prominent nucleoli. The background is fibrillary, composed of neural and fibroblastic tissues.



Figure 1b: Tumour bed after excision of the tumour.

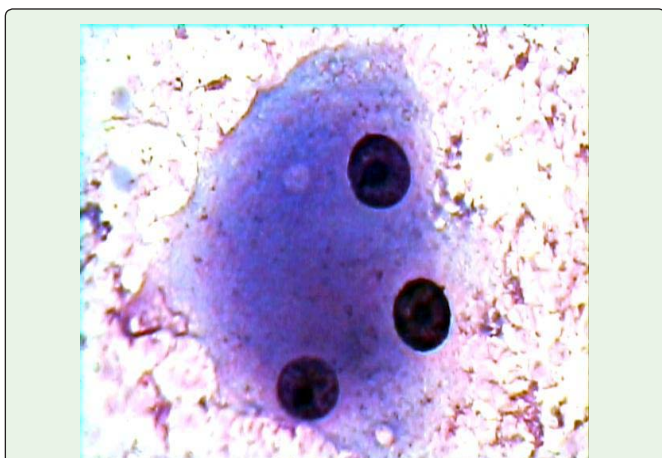


Figure 2: Cytology: MGG stain x 400. Large cells with basophilic cytoplasm with occasional multinucleation, s/o salivary neoplasm.

mature ganglion cells. However in 25% of ganglioneuromas elements of immature neurogenic tumors can be seen. It is hypothesized that neuroblastic tumors undergo a maturational process or spontaneous regression of neuroblastoma to ganglioneuroma [1]. Macroscopically, ganglioneuroma may appear encapsulated, although a true capsule is infrequent [2]. The tumour in our case was also well encapsulated and the decision to excise the mass was taken because it was found to be mobile, non-tender and there was no evidence of compression of any other structure.

Although ganglioneuroma is a benign tumour per se, there have been rare reports of metastatic ganglioneuroma. It is believed that these tumors represent metastases of neuroblastoma or ganglioneuroblastoma that have subsequently matured to ganglioneuroma; these patients have an excellent prognosis [4].

Treatment of cervical ganglioneuroma is surgical excision not only to confirm the diagnosis but also to prevent further growth and compression of neighboring structures. These tumors are not aggressive, and even in the case of incomplete resection, residual ganglioneuroma will not regrow or produce symptoms [3]. Injury to related neural and vascular structures during surgery may result in significant morbidity, occasionally leading to Horner Syndrome. However these symptoms usually resolve soon [1].

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