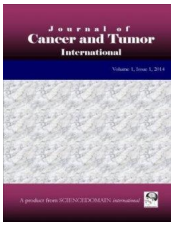

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Ganglioneuroma Involving Deep Facial Structures - A Case Report

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Authors' contributions

This work was carried out in collaboration between all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Ganglioneuroma is a rare peripheral neuroblastic tumor originating from the neural crest. It is a benign tumor involving posterior mediastinum and retroperitoneum. We present a case of 12-year-old boy who was diagnosed with ganglioneuroma involving deep facial structures - infratemporal fossa extending to adjacent structures.

Keywords: Ganglioneuroma; deep face; infratemporal fossa.

1. INTRODUCTION

Ganglioneuroma (GN) is an uncommon benign entity of the peripheral neuroblastic tumors (PNTs) that occurs commonly in childhood [1]. It arises from cells in the sympathetic ganglia and adrenal medulla. It is a fully differentiated tumor containing mature tissues. It is rare compared with other benign neural tumors such as schwannoma and neurofibroma but outnumbers neuroblastomas (NB) along the sympathetic axis by about 3 to 1. It is most often located in the posterior mediastinum, retroperitoneum and uncommonly in adrenal proper [2]. We present a 12-year-old boy who presented with the prominence of left cheek and proptosis of left eye

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and was diagnosed as GN of infratemporal fossa (IF) extending to adjacent structures.

2. CASE REPORT

A 12-year-old boy presented with the prominence of left cheek and bulging of the left eye of 3 months duration without any visual disturbance. On physical examination, he was able to fix and follow in both eyes with 2-3 mm of left eye proptosis but intact extraocular movement. Biomicroscopy revealed normal anterior segment with a regular and reactive pupil. His fundus and optic nerve examination were unremarkable. Hematological and biochemical investigations were normal. Vanillyl mandelic acid (VML) level was normal (4 milligrams / 24 hours). Computerised tomography (CT) of the brain, orbit, paranasal sinus and face followed by magnetic resonance imaging (MRI) revealed soft tissue mass involving IF extending to pterygopalatine fossa via pterygomaxillary fissure, sphenopalatine foramen and left orbit leading to proptosis. Intracranially it extended to apical part of the temporal lobe. Opacification of left maxillary sinus and ethmoid air cells was noted (Fig. 1). CT scan of chest, abdomen and pelvis were normal. Surgical resection of tumor was performed. On gross examination the excised mass measured 14 x 12x 10 mm. Histopathological examination revealed scattered nests of mature ganglion cells with distinct cell borders, single eccentric nucleus, prominent nucleolus, eosinophilic cytoplasm admixed with fascicles of Schwann cells. Stroma was densely collagenised with no involvement of bony trabeculae. There was no necrosis, atypia or mitosis (Fig. 2). Immunohistochemical staining was positive for S-100 in ganglia and Schwann cells (Fig. 3) and a diagnosis of GN was reached. Postoperatively the patient had an uneventful course.

3. DISCUSSION

GN is a rare PNT originating from neural crest which also includes the malignant histotypes NB and ganglioneuroblastoma (GNB) nodular and stroma-rich intermixed [1]. These tumors represent a spectrum of maturation from the most primitive form i.e. NB to the most mature form i.e. GN [3]. Robertson in 1915 defined GNB as a transitional tumor of sympathetic cell origin containing malignant neuroblastomatous and

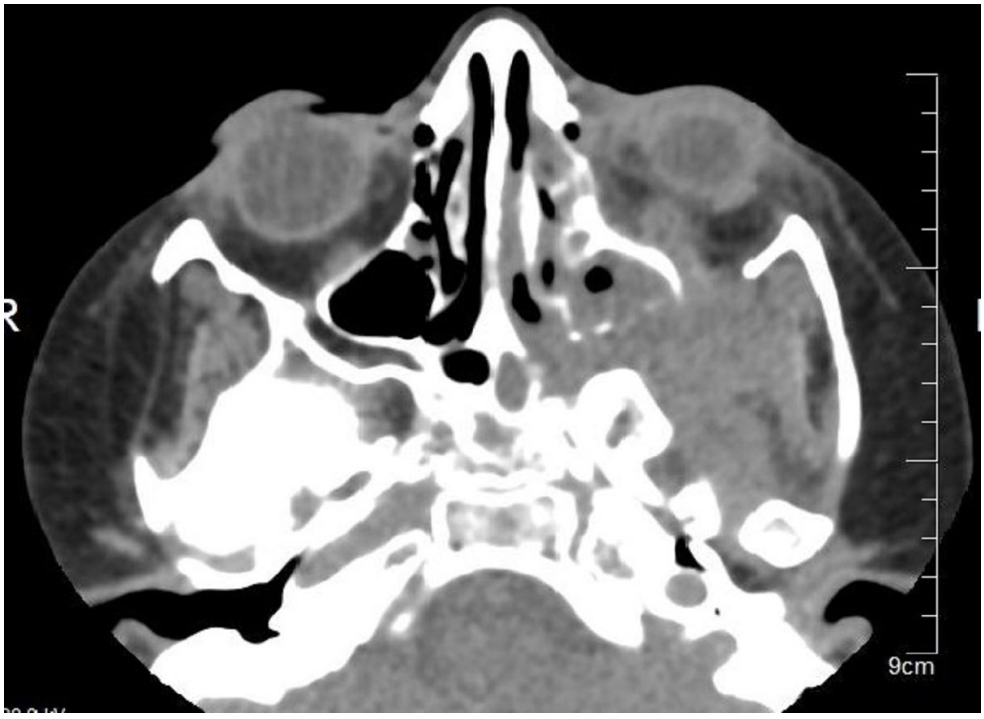

Fig. 1. CT scan image shows a soft tissue mass in the left infratemporal fossa extending to pterygopalatine fossa via pterygomaxillaryfissure, sphenopalatine foramen and to left orbit

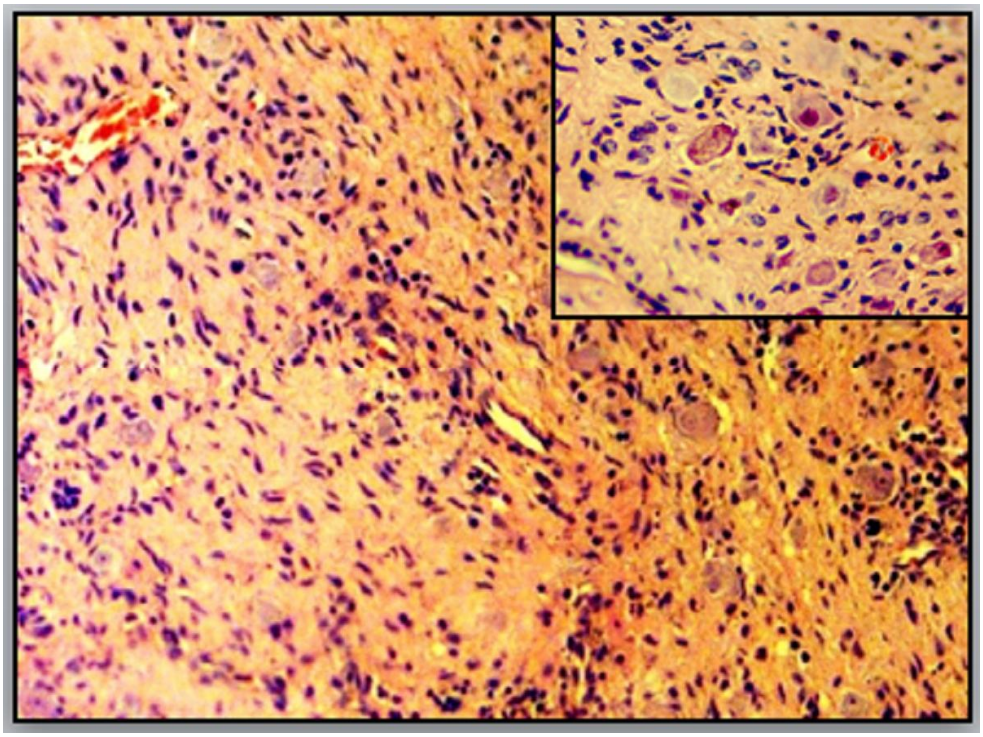

Fig. 2. Scattered nests of mature ganglion cells admixed with fascicles of spindle shaped Schwann cells and densely collagenised stroma. Mature ganglion cells with distinct cell borders, single eccentric nucleus, prominent nucleolus and eosinophilic cytoplasm seen (Hematoxylin and Eosin- 20x, Inset-40x)

benign ganglioneuromatous element [4]. There was no clear delineation between NB and GNB and between GNB and GN. The International Neuroblastoma Pathology Classification (INPC) in 1999 defined GN as a schwannian stroma-dominant tumor predominantly composed of ganglioneuromatous stroma with a minor component of scattered collections of differentiating neuroblasts and mature ganglion cells. GNB was characterized by well-defined microscopic nests of neuroblastic cells in various stages of differentiation, intermixed or randomly distributed in a ganglioneuromatous tissue in a background of abundant neuropil [1]. INPC in 2003 revision proposed four tumor categories in two distinct prognostic groups: favorable histology (FH) and unfavorable histology (UH) [5,6]. The INPC was the first to define the tumor categories using histologic indicators of both grade of neuroblastic differentiation and schwannian stromal development by accommodating the system developed by Shimada [7]. GN are classified into the FH group.

GN may arise denovo or by spontaneous or treatment-induced differentiation of NB or GNB. Most of the GN evolved through differentiating NB and this is supported by the findings of metabolically active GN (metaiodobenzylguanidine uptake, along with excretion of homovanillic acid and VMA), the higher age at diagnosis and higher percentage of immature GNs. It remains unclear which NB had the biologic potential to differentiate to GN, but it should be supposed that only NB exhibiting intact chromosomes 1, lack of MYCN amplification and near-triploid DNA values can mature into secondary benign ganglioneuromatous tumors [8]. The difference in distribution of NB and GN support the idea that most GN develop de novo rather than by way of maturation in a preexisting NB [9].

GN is more common in adolescents and young adults whereas NB and GNB mostly occur in infants and children [10]. In a study by Geoerger and colleagues the median age at diagnosis was 79 months (compared to 16 months for patients with NB). Males and females were equally affected. The commonest sites of origin were thoracic cavity (41.5%), abdomen excluding adrenal gland (37.5%), adrenal gland (21%) [9].

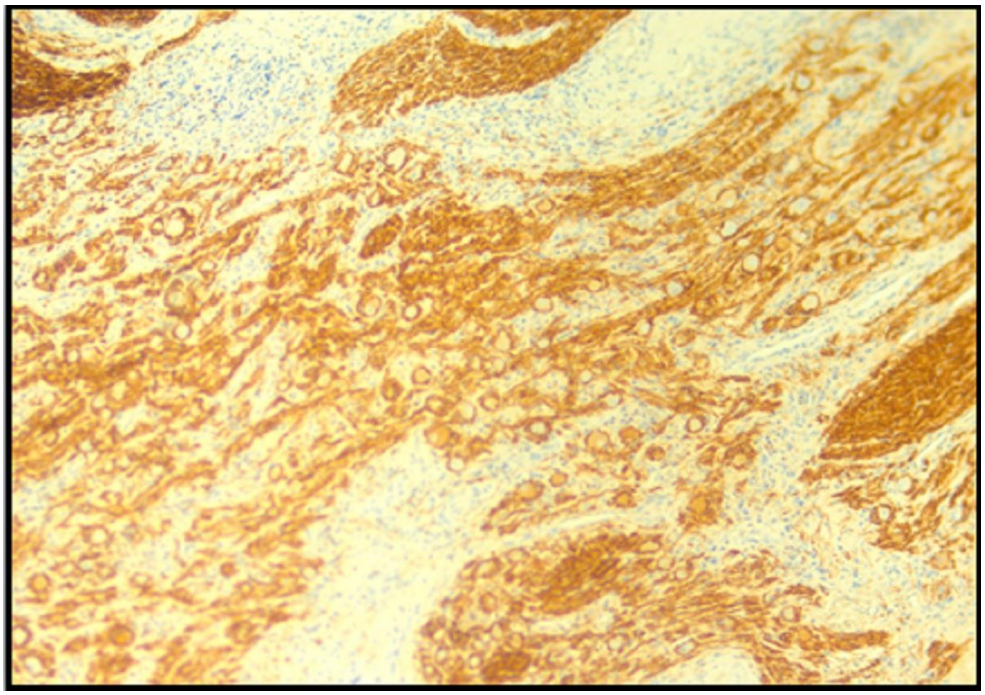

Fig. 3. Immunohistochemical study showing S100 positivity in ganglia and Schwann cells (S100x20x)

GN may also be found at other sites including the skin, retropharyngeal space, parapharynx, gastrointestinal tract (GIT), para-testicular region. In the GIT polypoid GN have been reported in association with several inherited diseases including Cowden syndrome, tuberous sclerosis, juvenile polyposis, type 1 neurofibromatosis and multiple endocrine neoplasia type IIb [2]. There were incidences involving head and neck but involvement of infratemporal fossa was not seen in any case. But our patient presented with GN involving IF and the tumor extended to pterygopalatine fossa via pterygomaxillary fissure, sphenopalatine foramen and left orbit which is a very rare presentation. The IF is a complex space that lies posterolateral to the maxillary sinus and many important nerves and vessels traverse it. These tumors usually come to clinical attention either incidentally or due to local mass effect on nerves [11]. GN may present with metabolic activity such as increased secretion of catecholamines and/or mIBG uptake. There are no specific diagnostic signs or symptoms discriminating GN and NB. Therefore GN requires tissue investigation for diagnosis.

Grossly the GN is a well-circumscribed tumor with a fibrous capsule. Histopathological examination (HPE) shows bundles of Schwann cells and mature ganglion cells [2]. HPE in our patient revealed scattered nests of mature ganglion cells admixed with fascicles of Schwann cells and densely collagenised stroma. On CT imaging, these tumors may be homogeneous or heterogeneous masses with low to intermediate attenuation. Calcification seen in 20% of cases is usually punctate as opposed to the coarse pattern seen in its malignant counterparts. Following contrast administration, the tumors demonstrate mild to moderate enhancement. On MR imaging GN appear as homogeneous mass with low and intermediate signal intensity on T1-weighted images [12]. In our patient MR signals were mainly of low intensity on T1 weighted images and of high intensity on T2 weighted images. Radiologically the differentials were lymphoma, Langerhans cell histiocytosis and sarcoma. Rare GN undergo malignant transformation and most commonly the malignant component resembles a malignant peripheral nerve sheath tumor [2].

Treatment for symptomatic extracranial GN consists of complete surgical excision, with radiological surveillance for local recurrence .In a study of 28 patients with complete resection and 12 with clinical tumor residuals no tumor progression was noted after treatment was completed [9].

4. CONCLUSION

GN are rare indolent benign neuroblastic tumors which are diagnosed histologically. Thorough sampling of the tumor is required to look for a neuroblastic component which will modify the diagnosis. They have an excellent prognosis after surgical resection.

CONSENT

As per international standard or university standard, patient's consent has been collected and preserved by the authors.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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