Case study

Ganglioneuroma of Deep Face- A case report

Abstract

Ganglioneuroma is a rare tumor of neural crest origin and comes under category of peripheral neuroblastic tumors .It is benign with most cases found in posterior mediastinum and retroperitoneum. Here we present a case of 12 year old boy who was diagnosed as ganglioneuroma of deep face with involvement of infratemporal fossa extending to adjacent structures.

Keywords:Ganglioneuroma ,deep face, infratemporal fossa

Introduction

Ganglioneuroma (GN) is the uncommon benign member of the peripheral neuroblastic tumors (PNTs) that occur commonly in childhood[1]. It arises from cells found in the sympathetic ganglia and the adrenal medula . It is fully differentiated tumor that contains no immature elements. 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 ,uncommonly in adrenal proper [2].We present a 12 year old boy who presented with prominence of left cheek and proptosis of left eye and was diagnosed as GN of infratemporal fossa extending to adjacent structures.

Case Report

A 12 year old boy presented with prominence of left cheek and bulging of left eye of 3 months duration without difficulty in seeing. On physical examination he was able to fix and follow in both eyes with 2-3 mm of left eye proptosis but full extra ocular movement. On biomicroscopy normal anterior segment examination with regular and reactive pupil was seen. His fundus and optic nerve examination were unremarkable. Hematological and biochemical investigations were normal. Vanillylmandelic acid: 4 milligrams /24 hours. Computerised tomography(CT) of brain, orbit , paranasal sinus and face and magnetic resonance imaging (MRI) revealed soft tissue mass involving infratemporal fossa extending to pterygopalatine fossa via pterygomaxillary fissure , sphenopalatine foramen and to left orbit leading to proptosis. Intracranially it extended to apical part of temporal lobe. Opacification of left maxillary sinus and ethmoid air cells were noted. Abdominal ultrasound, 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 were no features of necrosis , atypia or mitosis (Figure 2). Immunohistochemical staining was positive for S-100 in ganglia and Schwann cells (Figure 3) and a diagnosis of GN was made. Postoperatively the patient had a smooth course.

Discussion

GN is a rare tumor of neural crest origin and comes under category of the PNTs which also includes the malignant histotypes NB and ganglioneuroblastoma (GNB) nodular and stroma-rich intermixed[1] These tumor categories represent a spectrum of maturation from the most primitive form, NB to the most mature form, GN[3]. GNB was defined as a transitional tumor of sympathetic cell origin that contained malignant neuroblastomatous and benign ganglioneuromatous element by Robertson in 1915 [4]. There was no clear delineation between NB and GNB and between GNB and GN. The International Neuroblastoma Pathology Classification (INPC) established 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/or maturing or 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] .A revision in 2003 proposed four tumor categories in two distinct prognostic groups; that is, 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 always classified into the FH group. GN may evolve as a mature tumor from the very beginning or by spontaneous or treatment-induced differentiating NB or GNB. Intact 1p chromosomes are important for self-limiting behavior of NB. [8] Metabolic activity metaiodobenzylguanidine (mIBG) uptake, Homovanillic acid (HVA) and VMA excretion in a substantial fraction of patients with histologically proven GN, the higher age at diagnosis, and higher percentage of immature GNs suggest that most of the GN de facto evolved through differentiating NB. 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 . 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 of 49 patients with this tumor, the median age at diagnosis was 79 months (compared to 16 months for patients with neuroblastoma). Males and females were equally affected.. 41.5% arose in the thoracic cavity, followed by 37.5% in the abdomen outside of the adrenal gland; 21% arose within the adrenal parenchyma[9]. GN may also be found at other sites including the skin, retro- or parapharynx, the para-testicular region and gastrointestinal tract(GIT). In the GIT polypoid GN have been reported in association with several inherited diseases including Cowden syndrome, tuberous sclerosis and juvenile polyposis. Ganglioneuromatous polyposis has been described in patients with type 1neurofibromatosis and multiple endocrine neoplasia type IIb [2]. Our patient presented with GN of deep face involving infratemporal fossa extending to pterygopalatine fossa via pterygomaxillary fissure , sphenopalatine foramen and to left orbit which is a very rare location for this tumor. The infratemporal fossa 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 metaiodobenzylguanidine 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. Histologically it consists of

bundles of Schwann cells and mature ganglion cells [2]. 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 masses with low and intermediate signal intensity on T1-weighted images[12]. 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].

Conclusion

GN are rare indolent benign neuroblastic tumors which are diagnosed histologically. Thorough sampling of the tumor is required to look for neuroblastic component which will change the diagnosis and prognosis.They have excellent prognosis if surgically resected.

References

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Figure Legends

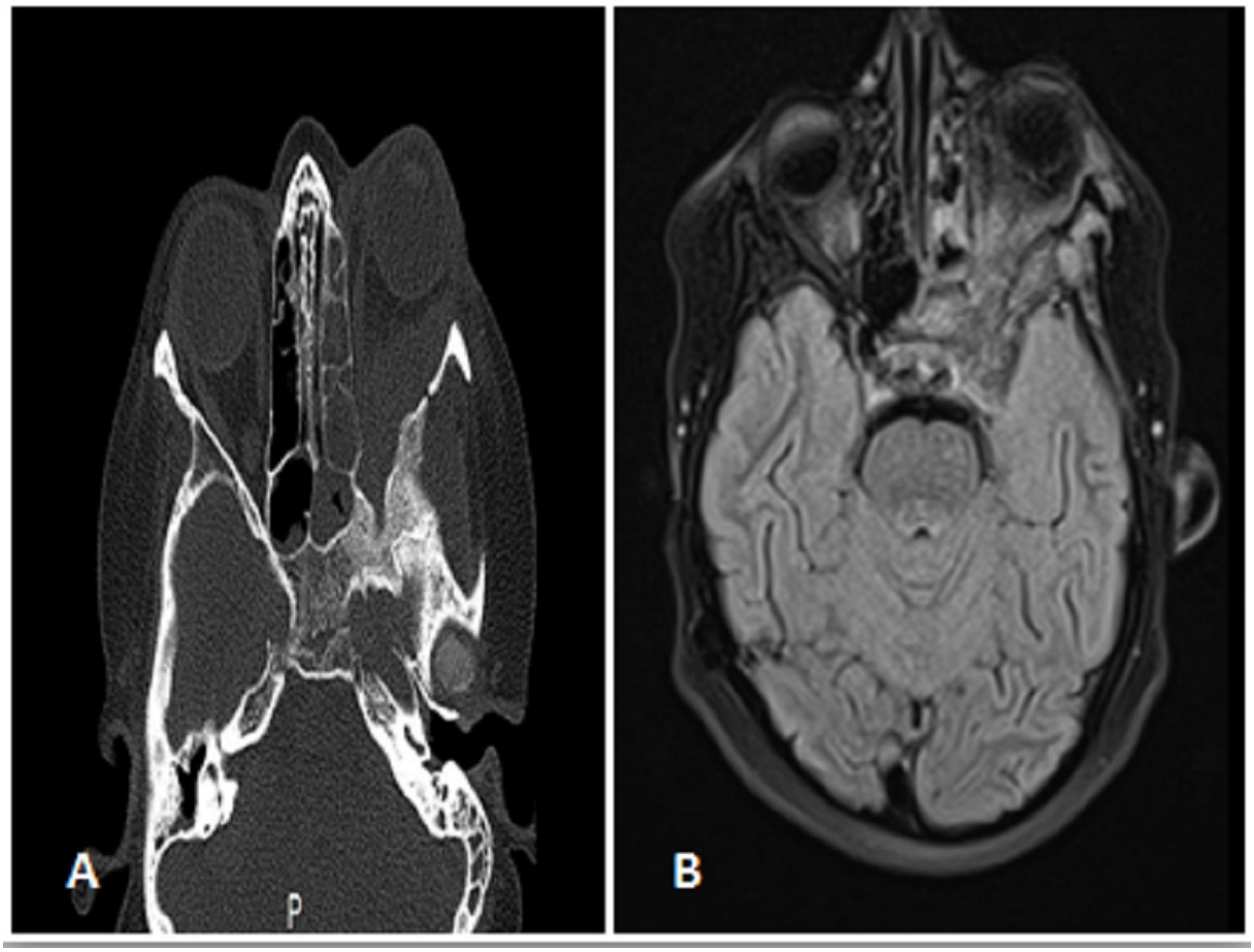

Figure 1.CT scans showing soft tissue mass involving infratemporal fossa extending to pterygopalatine fossa via pterygomaxillary fissure , sphenopalatine foramen and to left orbit.

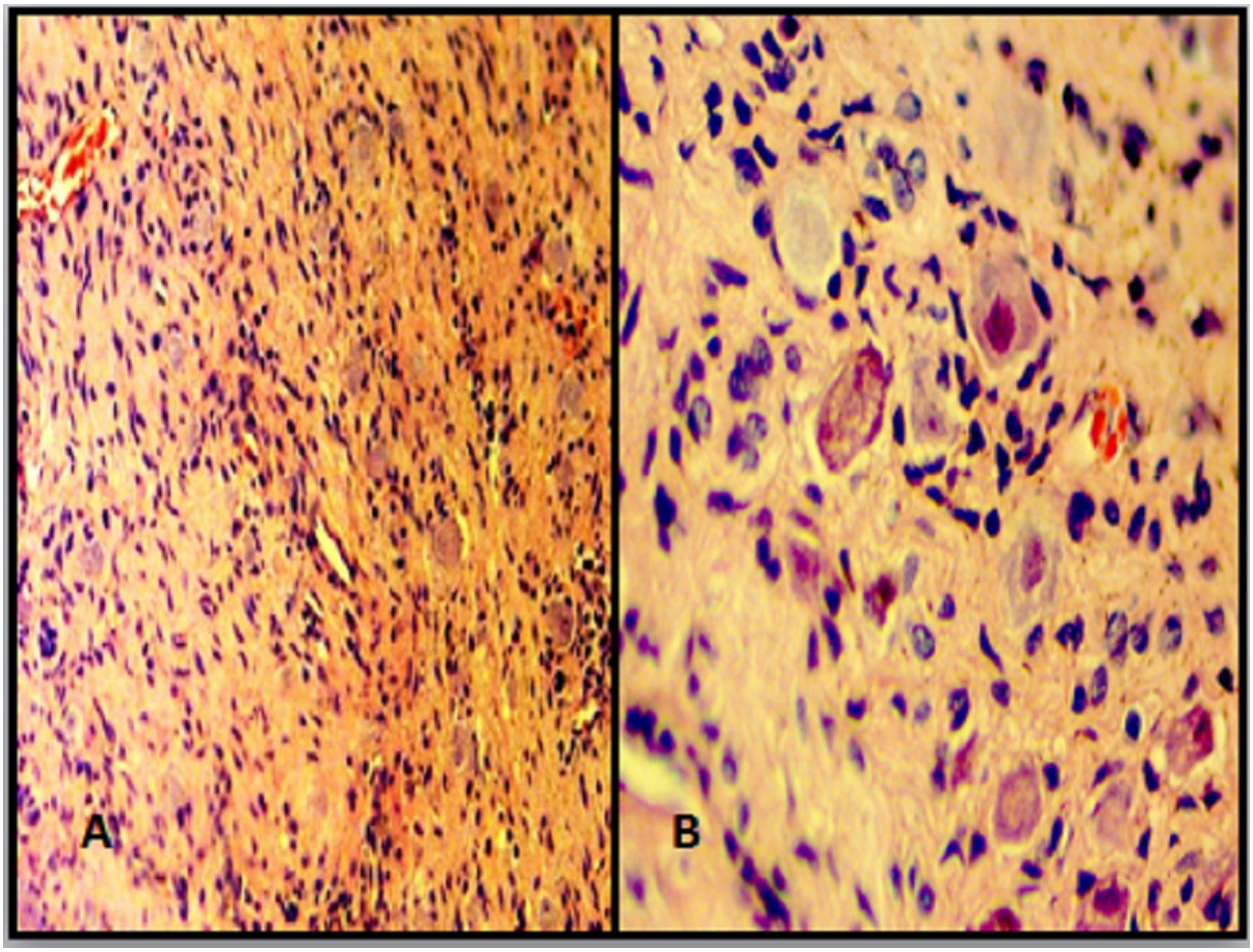

Figure 2. A. Scattered nests of mature ganglion cells admixed with fascicles of spindle shaped Schwann cells with densely collagenised stroma seen. B.Mature ganglion cells with distinct cell borders, single eccentric nucleus ,prominent nucleolus and eosinophilic cytoplasm (Hematoxylin and Eosin,A- 20X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

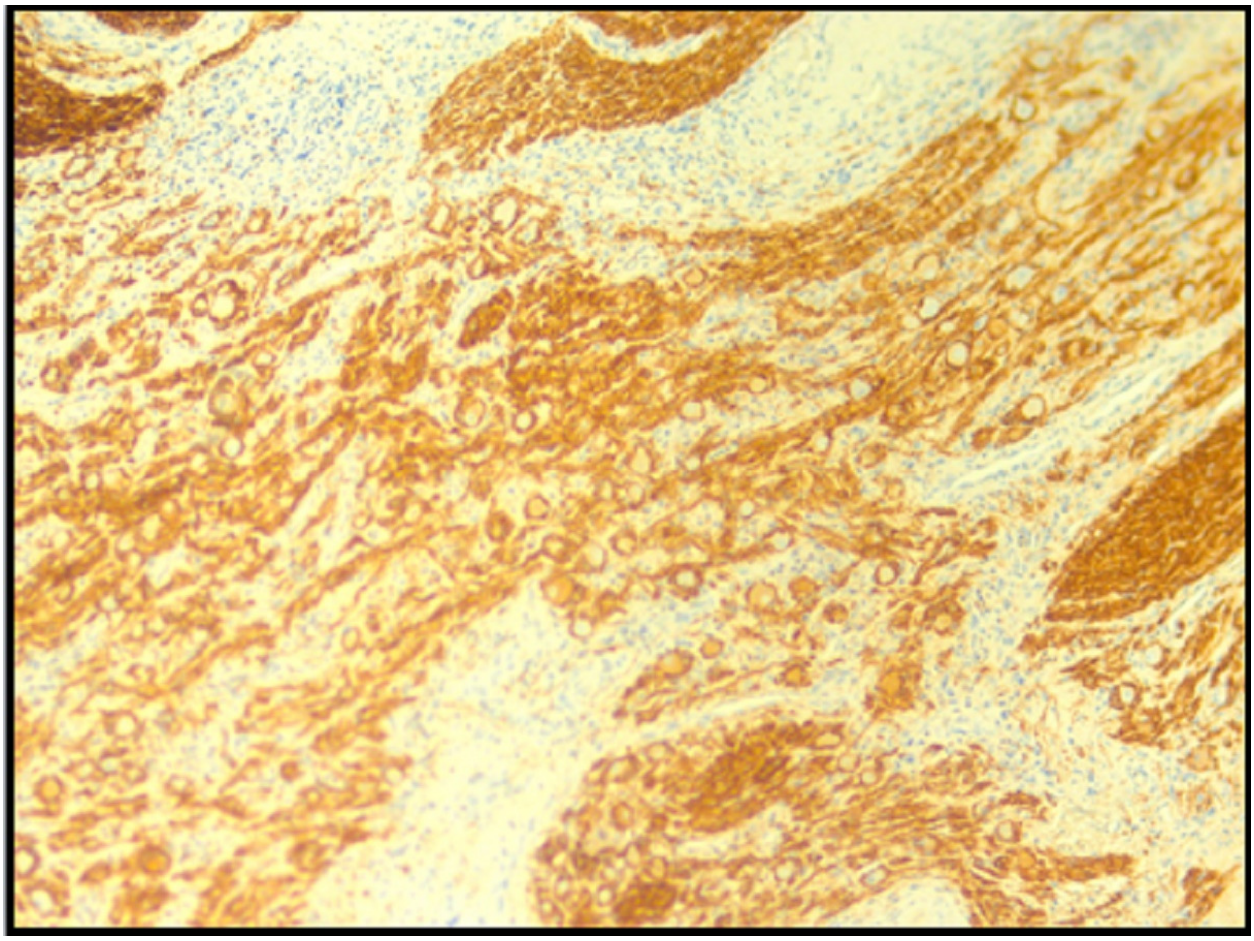

Figure 3 : Immunohistochemical study showing S100 positivity in ganglia and schwann cells(S100x20X)