Case Report

Surgical Management of Parapharyngeal Ganglioneuroma: Case Report and Review of the Literature

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Key Words

Head and neck neoplasms · Ganglioneuroma · Neurogenic tumor · Parapharyngeal space · Neck masses

Abstract

Parapharyngeal ganglioneuroma is a rare benign tumor, with fewer than 40 cases having been reported in the literature. We report a case of parapharyngeal ganglioneuroma in a child, including the presentation, diagnostic testing, treatment, outcome and a review of the literature. The patient presented with a large cervical mass arising from the cervical sympathetic chain. Complete excision of the ganglioneuroma was possible via a transcervical dissection approach without mandibulotomy. Clinical follow-up was conducted, and no recurrence has been observed to date.

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Introduction

Ganglioneuromas are benign tumors that arise from the peripheral derivatives of sympatheticoblasts, which are a cell line of neural crest cell origin [1]. Ganglioneuromas occurring in the head and neck region are rare, with fewer than 40 cases having been reported in the literature [2, 3]. Parapharyngeal space involvement is exceedingly rare [4].

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Fig. 1. **a** Coronal T1-weighted MRI demonstrating the mass extending superiorly to the level of the skull base. **b** Coronal STIR sequence revealing a well-encapsulated, lobulated mass extending posteriorly into the retropharyngeal space.

The symptoms of these tumors are usually related to the effects of the mass, nerve dysfunction or sympathetic activity due to secretory cells within the tumor [5]. Imaging techniques may be helpful for diagnosis; however, a biopsy is necessary for definitive diagnosis [6]. The most characteristic histological feature of ganglioneuromas is the presence of mature ganglion cells. In addition, they have no metastatic potential [7]. Surgical excision is the treatment of choice, and complete extirpation of the tumor is always recommended [8].

We present the case of a 5-year-old girl with a history of an enlarging cervical mass who underwent surgical resection of a parapharyngeal ganglioneuroma.

Case Report

A 5-year-old girl with a 4-month history of an enlarging left cervical mass presented to her pediatrician for evaluation. The mass was painless and was not associated with any systemic symptoms or local symptoms of obstruction. The patient was initially treated with a course of antibiotics without improvement. The mass continued to grow; therefore, the patient was transferred to our institution for investigation and definitive treatment. The MRI scan of the neck with contrast demonstrated an enhanced mass extending superiorly to the level of the skull base and posteriorly into the retropharyngeal space. There was anterolateral displacement of the common carotid artery and internal jugular vein. The lesion had increased signal intensity in the T2-weighted image and an isointense signal in the T1-weighted image. The STIR sequence revealed a well-encapsulated, lobulated mass with intense impregnation of the contrast material (fig. 1). The preserved planes of the soft tissues surrounding the tumor suggested that the lesion was noninvasive.

The patient underwent surgical excision of the mass via a transcervical dissection approach without mandibulotomy. Raising the skin flaps revealed reactive adenopathy of the level II and III lymph nodes. The internal jugular vein and the common carotid artery were observed to be displaced laterally due to the tumor mass. The tumor was found arising from the sympathetic chain, with the sympathetic chain ramifying into the mass. With careful sharp and blunt dissection, the anterior, posterior, and medial margins of the mass were dissected from the contents of the neck. The sympathetic trunk was clipped and divided at the point at which it ramified into the mass. All of the cranial nerves were identified and preserved. The mass was later freed from the surrounding tissues at the base of the skull and removed in toto (fig. 2).

Gross pathology revealed a lobulated, encapsulated mass of pink-tan soft tissue measuring $6.4 \times 4.5 \times 2.8$ cm in diameter. Microscopically, the mass was composed of mature ganglion cells, variable in number per field, with a compact eosinophilic cytoplasm, distinct cell borders and an eccentric and prominent nucleus. The stroma consisted of Schwann cells and mature fibrous tissue. Immunohistochemical reactivity for S-100 protein and synaptophysin was positive, confirming the diagnosis of ganglioneuroma (fig. 3).

Fig. 2. **a** Surgical field showing the relationship of the tumor to the carotid arteries. **b** Digital dissection of the tumor deep in the parapharyngeal space. **c** Surgical field after removal of the tumor showing the intact carotid bifurcation, internal jugular vein and adjacent cranial nerves.

Fig. 3. Schwannian cells with positive immunohistochemical reactivity for S-100 protein are shown (magnification $\times 300$).

The patient was extubated in the operating room with no clinical signs of respiratory distress. All of the cranial nerves were normal in the immediate postoperative course. The patient experienced an expected Horner's syndrome from division of the sympathetic chain and mild dysphagia, with aspiration of nonviscous liquids. On postoperative day 5, she was discharged for outpatient speech therapy to restore pharyngeal muscle function. Clinical follow-up was conducted, indicating that pharyngeal muscle function was restored in 2 months. Recurrence has not been observed to date.

Discussion

Tumors of the parapharyngeal space are rare. Up to 80% of these tumors are benign. Salivary gland tumors are slightly more common (38–48%) in the retropharyngeal and parapharyngeal space than neurogenic tumors (32–40%) [9]. The group of benign neurogenic tumors of the neck comprises neuromas (pseudotumors), neurilemmomas, neurofibromas, paragangliomas and the extremely rare ganglioneuromas [10].

Ganglioneuromas were first described by Loretz in 1870 and first reported as occurring in the neck by de Quervain in 1899. Sixty percent of patients with ganglioneuromas are under 20 years of age; the average age of presentation is approximately 11 years with a slight preponderance of female cases [4]. The hypotheses for the pathogenesis of benign ganglioneuromas include the spontaneously or artificially induced maturation of neuroblasts in a neuroblastoma into distinct ganglion cells, separation of the remaining cells from the

embryonic neural crest and necrosis of neuroblasts at an early stage of tumor development [11].

The development of ganglioneuroma is characteristically silent, and the appearance of clinical symptoms depends on the tumor size and site. These tumors most often manifest as an asymptomatic mass originating from the cervical sympathetic chain; other sites include the larynx, pharynx and the nodose ganglion of the vagus nerve [12]. Ganglioneuromas are considered to have a low level of secretory activity or no secretory activity, but even in the case of activity, patients are rarely symptomatic [4]. In the literature reviewed for this study, none of the parapharyngeal space tumors was reported to be metabolically active. In our case, the patient presented with an enlarging left cervical mass originating from the cervical sympathetic chain that did not have secretory activity.

A precise preoperative histological diagnosis of ganglioneuroma is difficult. A variety of methods are available. Fine-needle aspiration cytology as a diagnostic procedure is not always conclusive for ganglioneuroma. When performed preoperatively, it failed to identify the tumor in 60% of cases. A catecholaminergic crisis has never been described subsequent to fine-needle aspiration of a neck mass, but this theoretical possibility exists [13]. The value of a preoperative incisional biopsy is uncertain because although it may provide a conclusive diagnosis, the tissue planes will be obliterated, and unforeseen metabolic complications may occur.

The most characteristic histologic feature of ganglioneuromas is the presence of mature ganglion cells. Macroscopically, ganglioneuromas may appear to be encapsulated, although a true capsule is infrequent. Microscopically, the tumors are composed of intersecting bundles of spindle cells resembling neurofibromas or neurilemmomas, loose myxoid stroma and dysplastic ganglion cells that resemble normal ganglion cells both morphologically and immunohistochemically. Immunohistochemistry can be helpful in ascertaining the origin of the tumor and the differential diagnosis, but it is not essential for diagnosis [1]. Ganglion cells exhibiting positive immunohistochemical reactivity for S-100 protein are present in most cases. Malignant transformations of ganglioneuromas are exceptionally rare, possibly because growth regulation factors derived from Schwann cells have a protective influence on malignant tumor cells [14].

The current advanced imaging techniques may be useful for evaluating the extent of the tumor and differential diagnosis. CT most commonly reveals a well-encapsulated tumor with low attenuation and possible central densities. MRI demonstrates intermediate signal intensity and nonhomogeneous contrast enhancement [6]. Recent studies have shown that positron emission tomography could be helpful for early diagnosis of lesions of the parapharyngeal space and useful for surgical planning [15]. Ganglioneuroma is differentiated from salivary gland tumors in the parapharyngeal space by the presence of a fatty streak medial to the parotid gland, demarcating an anatomic boundary between the tumor and the gland [8]. In this context, the various other possible differential diagnoses for a cervical mass are tubercular adenitis, branchial cyst, lipoma, cystic hygroma, pharyngeal diverticulum, carotid body tumor, thyroid nodule and lymphoma [13]. Special attention should be paid to the rare cases of ganglioneuroma that are associated with multiple endocrine neoplasia type 2 and neurofibromatosis 1 [3].

Surgical excision is the treatment of choice. Numerous approaches for resection of neoplasms in the parapharyngeal space have been described, including the transoral, transcervical, transparotid, transcervical-transpharyngeal, and the infratemporal fossa approach and combinations of these. The location, size and pathological type determine the choice of the surgical approach [15]. In our patient, we performed a transcervical dissection without mandibulotomy.

Radiation treatment should be avoided because of uncertain results. Moreover, the young age of most patients may result in a possible growth retardation and the occurrence of late complications, such as the development of radiation-induced neoplasms [11]. The prognosis is generally favorable because ganglioneuromas lack metastatic potential [14].

Conclusions

Ganglioneuromas are rare childhood tumors that are usually first noticed as enlarging parapharyngeal or cervical masses. These tumors have the potential to secrete adrenergic products, although the tumor reported here did not. Ganglioneuromas are treated by surgical excision and, in most cases, can be readily removed transcervically without mandibulotomy. Both CT and MRI scanning offer anatomical information that may be useful at the time of surgery. Pathological characterization is necessary to confirm that the tumor is benign. Follow-up is suggested, both clinically and with repeat imaging if recurrence is suspected.

Disclosure Statement

The authors declare no conflicts of interest.

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