An Unusual Histology of Adult Cervical Ganglioneuroblastoma: A Case Report

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Abstract

Ganglioneuroblastic tumor is a primary malignant tumor of sympathetic chain, and can be found anywhere from the neck to retroperitoneum. It rarely occurs in the adult population. Here, we present a case of cervical ganglioneuroblastoma with a tiny neuroblastic component in a 33-year-old adult male. This case was diagnosed only by over sampling.

Keywords: Ganglioneuroblastoma; Neck; Over sampling

1. Introduction

Ganglioneuroblastoma is a primary malignant tumor of the sympathetic chain. This tumor is in the spectrum of neural crest derived tumors ranging from neuroblastoma at one end, ganglioneuroma at the other and ganglioneuroblastoma in between.

This tumor rarely occurs in adults and even more uncommon in the cervical region.

In this report, we present the case of an adult with large cervical ganglioneuroblastoma with tiny neuroblastic component, which was diagnosed by over sampling.

2. Case Report

A 33-year-old man was admitted to our hospital complaining about cervical swelling in right side in the past two months. The painless mass was insidious in onset and slow in progression. There were no abnormal findings in his laboratory data including blood and urinary catecholamine
concentrations, past medical history or family history.

On local examination, the mass was mobile and extended from parotid tail to the supraclavicular region. Sensory or motor disorder of facial nerve was not seen. In the oropharyngeal examination, right tonsil hypertrophy and shifting to the midline were encountered.

CT scan of the neck was conducted, which revealed a single discrete lump (6x1 cm) in the right side, posterior to the sternocleidomastoid muscle. (Fig. 1)

En masse excision was done following exposure of vessels and nerve. It was sent for histopathological study.

In gross evaluation, it was an encapsulated mass with solid glistening yellow cut surface and soft consistency. (Fig. 2) In microscopic examination, proliferation of admixture of Schwann cells with focal accumulation of mature as well as maturing ganglion cells was revealed (Fig. 3A)

There was a small microscopic nest (0.6 mm) of immature neuroblastic cells with high N/C ratio and rosette formation. (Fig. 3B) Immunohistochemistry study was performed for GFAP marker and positive staining was observed in Schwann cells and ganglion cells.

Eleven months after surgery, the patient was asymptomatic, and no recurrence was detected.

**Fig. 1.** Cervical CT scan revealed a lump posterior to SCM muscle.
3. Discussion

Ganglioneuroblastomas (GNB) are rare childhood neoplasms. They can also appear in adults, but it occurs less frequently. More than 90% of cases are reported in children younger than five. In one series study of 80 cases of GNB, only three patients were above 20. According to Yamanaka et al., only 33 cases of retroperitoneal GNB in adults have been noted in English medical literature. Mehta et al. (1997) in their study have reported a case of bilateral intra-abdominal GNB in a patient of age 20 among the Indian population.

In an article, Kilton et al. (1976) published a review of 33 cases of GNB in adults worldwide. GNBs
occur wherever sympathetic nervous systems are present. Mediastinum and retroperitoneal tissues are the most common sites for ganglioneuroblastomas. Reby et al. evaluated 15 adult patients with NB (neuroblastoma)/GNB. The overall average age of the patients at the time of diagnosis was 30. Primary tumor origins were in pelvis, mediastinum, abdomen, adrenal gland, retroperitoneum, and mixed locations.

In adults, GNB is generally discovered accidentally or by compression symptoms, as in the present case.

Preoperative diagnosis is difficult, and preoperative biopsy may produce results depending upon which component is biopsied. It may look like a small round cell tumor or ganglionic cell component in Schwannian stroma.

Horner syndrome is more frequently associated with para pharyngeal space tumors rather than sympathetic chain tumors. Furthermore, the nerve of origin is not always the sympathetic chain even in cases with Horner syndrome. But our case had no symptoms of neural involvement.

Sometimes, laboratory test results can be helpful in making a preoperative diagnosis. Fifty-seven percent of patients with a ganglioneuroblastoma show increased concentrations of serum serotonin and urinary catecholamine and their metabolites (vanil mandelic acid and homovanillic acid). When they are initially positive, hormonal examinations are reliable for the follow-up of recurrence in children. But our patient had no abnormality in his laboratory tests.

There are two subtypes described in existing literature: the nodular subtype includes gross nodules of neuroblastoma (small immature cells) in wide expanses of ganglioneuroma (large mature cells in a fusiform stroma), whereas the intermixed subtype consists of microscopic nests of neuroblastoma settled in a ganglioneuroma Tous stroma.

Curative treatment should be a complete resection. However, in case of only partial resection, adjuvant radiotherapy may be proposed with a close follow-up and regular re-evaluation. Surgical excision is often the most effective treatment. This treatment can be followed by radiotherapy only when continued tumor growth has occurred.

4. Conclusion

Cervical intermix Ganglioneuroblastoma is an extremely uncommon tumor in adults. A thorough sampling of neurogenic tumors is always critical for accurate diagnosis. Without wide sampling, the tumor in this study would have been diagnosed as ganglioneuroma. Surgical Pathologist should distinguish this tumor when dealing with adult cervical mass even though it is a rare entity.

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