

A CASE OF GANGLIONEUROMA*

Yavuz Özorán MD**, Şevket Ruacan MD***, Sezer Ş. Komşuoğlu MD****
Afet Özorán MD***** , Fadıl Aktürk MD*****

Key words: ganglioneuroma, von Recklinghausen's disease, neurofilaments, satellite cells

Isolated ganglioneuromas are rare tumors in the absence of von Recklinghausen's disease¹⁻³. There are pathologic diagnostic difficulties with the metastatic lesions of ganglioneuroblastomas and ganglioneuromas. Neuroendocrinologic manifestations of these two types of neoplasms are also controversial topics in diagnosis. Apart from these two important items, ganglioneuromatosis may also be seen in this condition; ganglioneuromas are a component of this syndrome³. The rarity and isolated presence of this tumor prompted us to present this case.

Case Report

An eight-year-old girl in good health was admitted to our hospital because of a nodule localized to the front of the left sternocleidomastoid muscle. She had been on non-specific antibiotic therapy for a month before her admission. Physical examination revealed a solid, mobile tumoral nodule measuring 3 x 2 x 1 cm in diameter located at the front of the left sternocleidomastoid muscle. No associated skin lesion was present. The other systems were within normal limits. Clinical or laboratory signs of tuberculosis were negative. Macroscopic appearance of the excisional biopsy material revealed a solid mass measuring 3 x 2 x 1 cm in diameter and off-white in color. Microscopically, the tumor revealed mature ganglion cells intermixed with fascicles of spindle cells. There was no satellite cell association (Fig. 1).

* From the Departments of Pathology, Karadeniz University Faculty of Medicine, Trabzon and Hacettepe University Faculty of Medicine, Ankara.

** Associate Professor of Pathology, Karadeniz University Faculty of Medicine.

*** Associate Professor of Pathology, Hacettepe University Faculty of Medicine.

**** Associate Professor of Neurology, Karadeniz University Faculty of Medicine.

***** Associate Professor of Histology and Embryology, Karadeniz University Faculty of Medicine.

***** Associate Professor of Neurosurgery, Karadeniz University Faculty of Medicine.

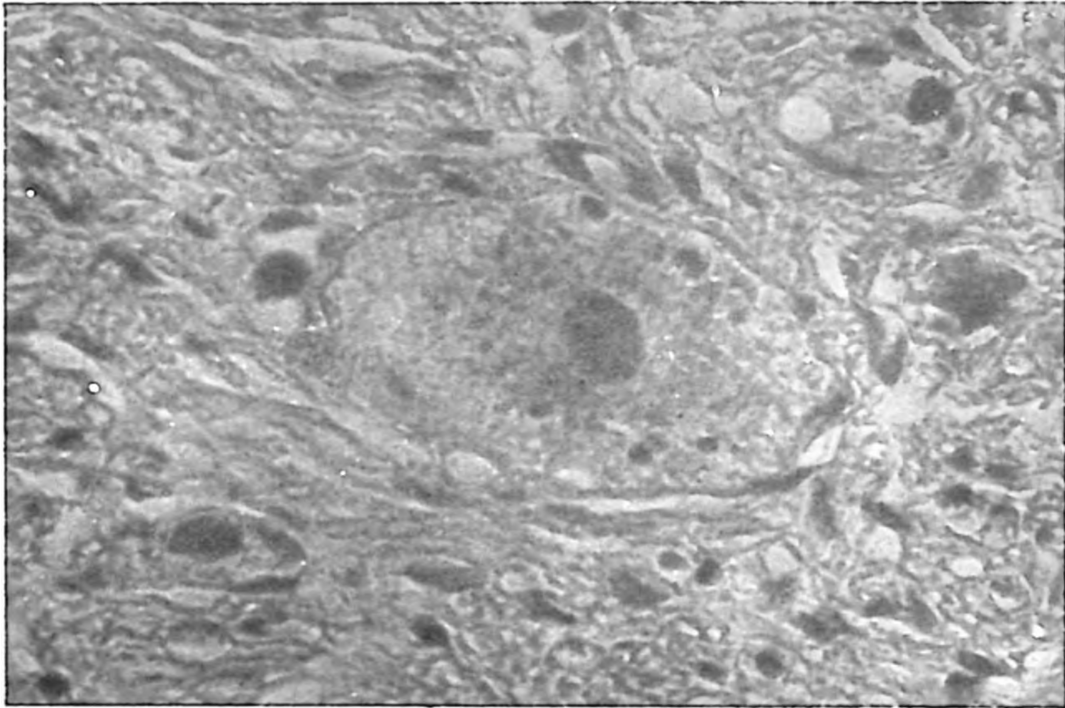


Fig. 1.: A ganglion cell is seen in the center intermixed with fascicles of spindle-shaped cells (HE x 40)

Discussion

Isolated ganglioneuromas are rare tumors which may be associated with von Recklinghausen's disease¹. They are in close proximity to the autonomic nervous system e.g. cervical or paravertebral, as in our case. The absence of satellite cell association is the most useful histologic finding that helps in differentiating this tumor from von Recklinghausen's disease, as evidenced in our case¹.

It is difficult to explain the isolated ganglioneuroma noted in our patient, who did not present with evidence of von Recklinghausen's disease or neuroblastoma. It may be that this tumor had been dormant since childhood. Immunohistochemical and ultrastructural studies prove that microfilaments which occur in ganglioneuromas are real neurofilaments^{2,4}. This finding confirms their hamartomatous neuronal origin. They may also be seen as a component of Cowden's disease³. In this situation, they occur in viscera apart from soft tissues³.

The absence of neuroendocrinological signs and systemic abnormalities confirmed that this patient had an isolated ganglioneuroma.

Summary

The case of an isolated ganglioneuroma localized to the cervical region in an eight-year-old girl is presented. The rarity of this disease in the absence of associated systemic and neuroendocrinological abnormalities is discussed.

REFERENCES

1. Geffner RE, Hassell CM. Ganglioneuroma of the skin. *Arch Dermatol* 122:377, 1986.
2. Mukai M, Torikata C, Iri H, et al. Expression of neurofilament triplet proteins in human neuronal tumors. *Am J Pathol* 122:28, 1986.
3. Lashner BA, Riddell RH, Winans CS. Ganglioneuromatosis of the colon and extensive glycogenic acanthosis in Cowden's disease. *Dig Dis Sci* 31:28, 1986.
4. Yokoyama M, Okada K, Tokue A, et al. Ultrastructural and biochemical study of benign ganglioneuroma. *Virchows Arch (Pathol Anat)* 361:195, 1973.