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Clinical Research

GANGLIONEUROBLASTOMA - A CASE REPORT

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ABSTRACT Neuroblastomas are among the most important tumors of extra cranial origin in pediatric patients. They arise from embryonal cells involved in the development of the sympathetic nervous system, whose differentiation has been arrested [1, 2].

This article describes an atypical presentation of ganglioneuroblastoma in a 52 year old male patient. X- Ray left hand in AP and Lateral views showed it as a soft tissue swelling. Excision was done under digital block, and excised specimen was sent to department of pathology for histo pathological examination. HPE showed it as gangioneuroblastoma and pathologists advised to get immunohistochemistry for cathepsin D to be done.

KEYWORDS : Ganglioneuroblastoma, Schwannian Stroma, Immunohistochemistry, Cathepsin D.

Introduction

Ganglioneuroblastoma is a rare childhood tumour and rarely appears in adults. In adults, this tumour is generally discovered by accident or by compression Neuroblastic tumors account for 8–10% of all tumors diagnosed in pediatric patients and 80% of those found in children less than 5 years of age [3]. The most common locations include the adrenal glands (35%), paraspinal retroperitoneal ganglia (30–35%), posterior mediastinum (20%), head and neck (1–5%), and the pelvis (2–3%) [4]. Imaging studies play fundamental roles in the diagnosis and follow-up. Biopsy excision biopsy and pathological study is the diagnostic. Treatment options include surgery, chemotherapy, and radiotherapy. New prospects are emerging with the use of biological therapy based on the use of immunomodulators and retinoids.

Review of Literature

Can J Surg 2009 Aug; 52(4): E111–E113. PMCID: PMC2724818

Nodular ganglioneuroblastoma in adults

Thierry Peycru, MD, Jérôme Guiramand, MD, Eric Tardat, MD, Pierre-Henri Savoie, MD, Jean-Phillipe Avaro, MD and Paul Balandraud, MD

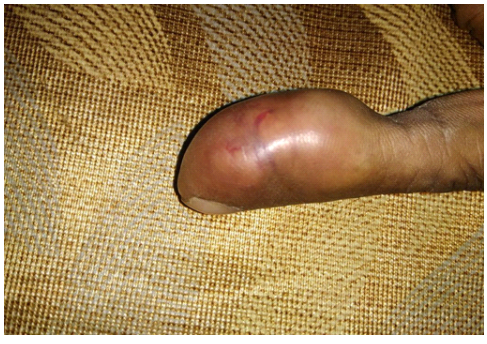
A 34 year-old otherwise healthy woman was admitted to hospital for atypical right-sided back pain that was radiating to her thigh. A computed tomography (CT) scan showed a 4-cm homogeneous retroperitoneal tumour without contrast enhancement .They performed a CT-guided biopsy. The pathological examination result was interpreted as a probable solitary fibrous tumour. Histological and immunohistochemical examination results confirmed the diagnosis of a nodular ganglioneuroblastoma.

Materials and Methods

Mr. Pentaiah, a 52 year old male patient came from kethireddypalli presented to hospital on 15/10/2017 with swelling in left middle finger for 4 months. It was painless. No history of any injury. Had past history of same swelling in the same place for 2 months for which he underwent excision on 24/03/2017 in this hospital.

On local examination of hand

- A 3 x 3 cm solitary swelling present on the palmar aspect of middle and distal phalynges of middle finger of left hand.
- A scar was present over the swelling.
- Swelling was oval in shape with ill defined borders.
- Surface is bosselated and irregular.
- Non tender and no local rise of temperature.
- Firm in consistency.
- Not fixed to bone. Joint movements were normal.



Funding: None

Study Procedure

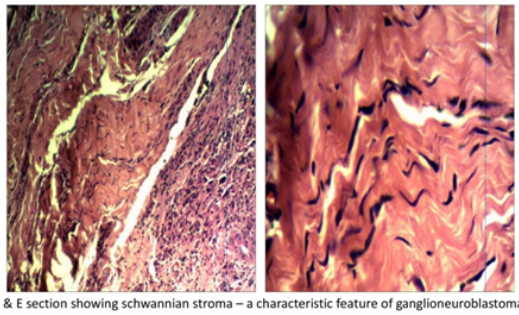
Surgical work up was done. X- Ray left hand in AP and Lateral views showed it as a soft tissue swelling. Excision was done under digital block, and excised specimen was sent to department of pathology for histo pathological examination.

Results

HPE showed it as gangioneuroblastoma and pathologists advised to get immunohistochemistry for cathepsin D to be done.

Immunohistochemistry

Neuroblasts: Neuron specific enolase, CD 57, CD 56, synaptophysin, chromogranin.Ganglion cells: S 100, synaptophysin, glial fibrillary acidic protein Schwannian stroma: S 100, synaptophysin Negative for Pancytokeratins; Epithelial membrane antigen.



H & E section showing schwannian stroma – a characteristic feature of ganglioneuroblastoma

Patient came again with recurrence of the swelling and got admitted on 4/1/2018 for surgery.

Discussion

Ganglioneuroblastoma is a rare childhood tumour and rarely appears in adults. In adults, this tumour is generally discovered by accident or by compression. It is a tumour of intermediate grade arising from sympathetic nervous tissues. Exhibit a degree of differentiation intermediate between neuroblastoma & ganglioneuroma. Predominant in younger age group but typical examples of adult cases are also not uncommon. Most common locations are the retro peritoneum & mediastinum. Firm in consistency, frequent calcifications. The clinical manifestations vary widely and are frequently nonspecific at the outset (asthenia, sweating, fever, pain related to the primary tumor or to metastatic lesions, and abdominal distension).

Differential Diagnosis

1. Implantation dermoid.
2. Infected cyst.
3. Synovial Sarcoma
4. Ganglion
5. Hamartoma
6. Neurofibroma
7. Desmoplastic small round cell tumour
8. Ewings sarcoma/PNETs
9. Ganglioneuroma
10. Lymphoma
11. Rhabdoid tumour
12. Melanoma
13. Neuroblastoma
14. Schwannoma with neuroblastoma like features

Pathological Examination

Gross: homogenous on cut section; cystic/hemorrhagic areas correlate well with the neuroblastic component.

Microscopy: 2 variants

- Schwannian stroma rich
- Schwannian stroma poor

Schwannian stroma rich/Imperfect

All stages of neuronal differentiation are seen. Collections of ganglion cells, many of them immature, multinucleated or otherwise abnormal. Fine, fibrillary cobwebby network is seen in between masses of cells – diagnostic feature – due to neurites emanating from the tumour cells.

Schwannian stroma poor/Immature

Gross & microscopic appearance of ganglioneuroma except for the presence of well defined highly cellular areas of neuroblastoma. Schwann cell elements are non-neoplastic. Neuroblastic foci can be confused with the foci of perivascular lymphocytes. Boundaries between the two tumours tend to be sharp.

Our study is comparable with the following study.

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They performed the resection via a transabdominal approach. The tumour was encapsulated and easy to remove except near the L2 paraspinal region, where dissection was difficult. There were no perioperative complications, including blood loss. They discharged the patient on the sixth postoperative day. Pathological examination revealed a lobulated circumscribed solid mass. Histological and immunohistochemical examination results confirmed the diagnosis of a nodular ganglioneuroblastoma.

The resection margins revealed microscopic residual tissue (R1) in the L2 paraspinal region. Ganglioneuroblastoma immunohistochemical patterns

Cell	Immunohistochemistry
Fusiform Schwann	Ps100+
Ganglioma and neuroblastoma	NSE+ Chromogranin+ Synaptophysin+ CD56 (NCAM)+

NCAM = neural cell adhesion molecule;
NSE = neuron-specific enolase.

Merits and Demerits

This study shows an atypical presentation of ganglioneuroblastoma which is rarely seen in adults. Recurrent swelling needs excision biopsy and diagnosis is made accordingly.

However very few studies are there regarding ganglioneuroblastoma and the present study was conducted up on patient who attended this institution and may not represent the etiology of swelling in the population.

Recommendations

In adults, this tumour is generally discovered by accident or by compression. Preoperative diagnosis is difficult, but x-ray or sometimes CT scan generally reveals a mass with smooth margins, and it is sometimes possible to discern a heterogeneous pattern, tumour calcifications and contrast enhancement. Characteristics of the ganglioneuroblastoma on MRI have been detailed elsewhere and can help to discriminate between an adenoma and a cyst.

It is also important to note that preoperative biopsies can produce false-negative results. Ganglioneuroblastomas can look like a neuroblastoma in a partial ganglioneuroma stroma. In the nodular subtype, the neuroblastoma nodules and ganglionic cells can represent less than 5% of the tissue. A large biopsy is required for the diagnosis.

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). [9] When they are initially positive, hormonal examinations is reliable for the follow-up of recurrence.

Conclusion

Ganglioneuroblastoma in adults is very uncommon. Characteristics of this tumour remain poorly understood, and preoperative diagnosis is difficult. Excision was done under digital block, and excised specimen was sent to department of pathology for histo pathological examination. HPE showed it as gangioneuroblastoma. 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.

In adults, prognosis depends on surgical margin resection. Adjuvant radiotherapy may be required.

- Definitely better than that of neuroblastoma.
- Worse for the immature variety than the Imperfect
- Worse if the neuroblastoma component is in the form of nodular aggregates.
- Same prognostic criteria as that of Neuroblastoma assessment are applicable as well.

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