http://jmscr.igmpublication.org/home/
ISSN (e)-2347-176x ISSN (p) 2455-0450
crossref DOI: https://dx.doi.org/10.18535/jmscr/v11i2.18

IGM Publication
Journal Of Medical Science And Clinical Research
An Official Publication Of IGM Publication

Ganglioneuroma of Coccyx: A Case Report

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Abstract
A child aged 5 years female, complained of lower back pain for 5 months during defecation. Per-rectal examination revealed a swelling in retrorectal presacral region. She was operated. A tumour, measuring 8×5×5 cm was excised. Histopathological examination of the tumour revealed presence of large number of ganglion cells and schwann cells in loose myxomatous fibrillary stroma. Tumour tissue revealed positivity for S100, desmin and neuron-specific enolase (NSE). In addition, mild positive reaction (1+) was obtained with anti-Sox-10 antibody. Negative reaction was obtained with anti-Ki 67 and anti-chromogranin antibodies. Tumour was finally diagnosed as ganglioneuroma.
Keywords: *Benign Ganglion cells, Schwann cells, S100 positivity.*

Case Report

A 5-year old female complained of dull lower back pain during defecation for last 5 months. Present case relates to a patient with soft tissue neural tumour epicentered in retrorectal presacral region. Patient was operated. Biopsy pieces were taken in two vials. First vial contained two pieces, together measuring 2×1.5×1 cms. Second vial contained a soft tissue tumour piece, measuring 8×5×5 cm. All the pieces were sectioned. Microscopic examination showed tumour tissue, comprising of large number of ganglion cells, neurites and schwann cells in diffuse loose myxomatous stroma. Ganglion cells had eosinophilic cytoplasm, single eccentric nuclei and prominent nucleoli. Mild perivascular and diffuse lymphocytic infiltration was also seen (figure 1). Immunohistochemistry revealed strong positivity (3+) of tumour cells with anti-S100 antibody. Moderate positivity was seen with anti-desmin and anti-neuron specific enolase antibodies. Moreover, mild positive reaction (1+) was obtained with anti-Sox-10 antibody. Negative reaction was obtained with anti-Ki 67 and anti-chromogranin antibodies (figure 2).

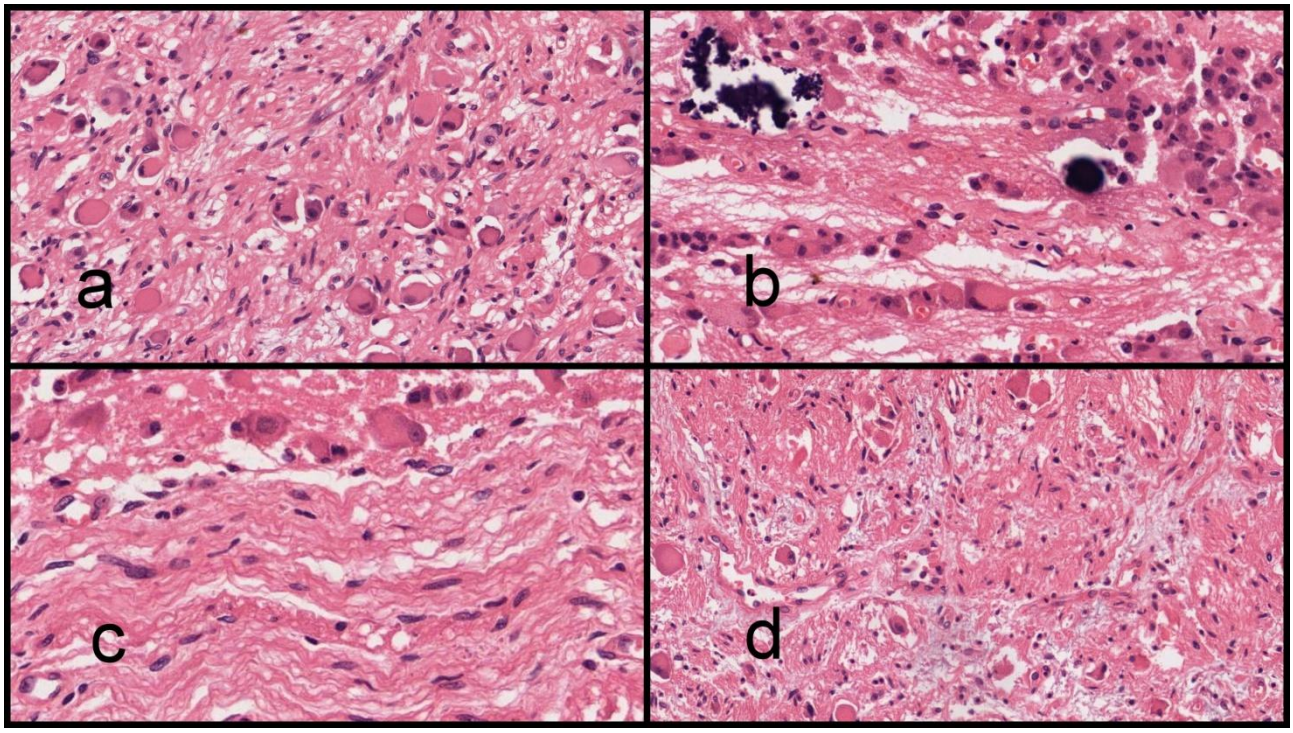

Figure 1 (a) Photomicrograph shows abnormal clustered ganglion cells. Ganglion cells had eosinophilic cytoplasm and single eccentric nuclei (HE×400). **(b)** Photomicrograph shows foci of calcification (HE×100). **(c)** Photomicrograph shows neurites with parallel arrangement of nuclei (HE×400). **(d)** Photomicrograph shows loose myxomatous stromal tissue (HE×100).

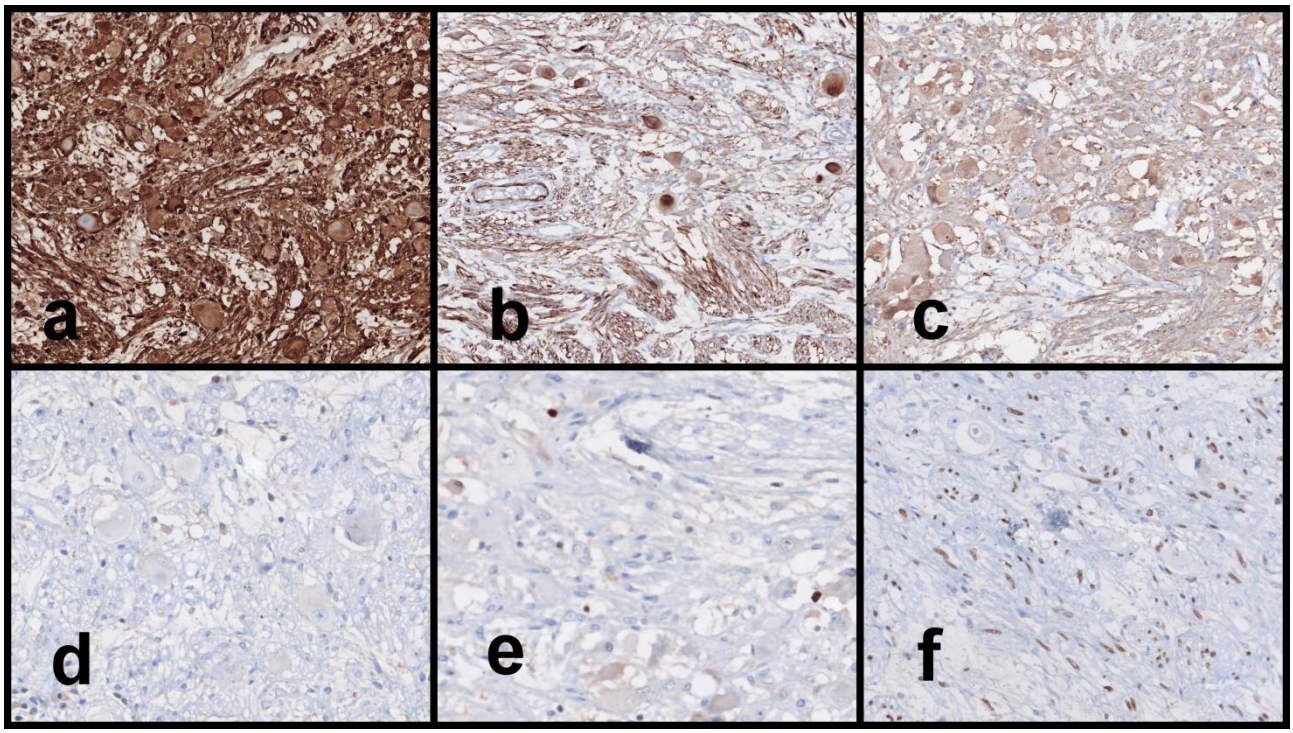

Figure 2 (a) shows strong positive (3+) staining of fusiform tumour cells for S100 antigen with anti-S100 antibody. **(b)** shows moderate positive staining (2+) of tumor cells with anti-desmin antibody. **(c)** shows moderate (2+) staining of tumor cells for neuron-specific enolase (NSE). **(d)** shows negative staining for Ki 67 antigen. **(e)** shows negative staining for chromogranin. **(f)** shows mild nuclear staining (1+) of schwann cells for Sox-10 antigen using anti-Sox-10 antibody.

Discussion

Ganglioneuromas (GN) are a benign, slow-growing neoplasms arising from one of the most mature neuroblast cells of the neural crest[1]. Tumour does not contain immature neuroblastic elements. It commonly develops in retroperitoneum[2] or posterior mediastinum[3,4,5]. Microscopically, ganglion cells and schwann cells are seen in myxoid fibrillary stroma. Neuron-specific enolase (NSE) or peptide hormones may be demonstrated in ganglion cells[1]. Further, one third of the patients may present with calcification in the tumour tissue. Similar metastatic calcification was seen in the current case. Clinically, the patient may present with sweating, hypertension and /or diarrhea due to release of catecholamines[4]. However, these clinical features were not found in the present case. Moreover, pigment (neuromelanin) may be found in the ganglion cells. Present case did not show pigment in ganglion cells[3,4]. Rarely, adipocytes may be found in tumour cells[2]. This feature was also not found in the present case. Similar tumour may also arise from the adrenal medulla[6]. One other patient had malignant peripheral nerve sheath tumour arising from adrenal ganglioneuroma[6]. Cutaneous ganglioneuroma is rare[4] and in a few instances, ganglion cells alone may be found in the tumour without schwann cells. This type of benign neoplasm is called ganglion cell choristoma[7,8]. Ganglioneuroma is a neoplasm of sympathetic nervous system. These tumours mainly occur in young adults and older children[1,9]. However, present tumour developed in a young female child directly from the mature ganglion cells[4]. Further, ganglioneuromas do not express chromosomal aberrations which are commonly found in neuroblastoma[1]. Rarely, ganglioneuroma may behave as a malignant neoplasm[4,5]. Moreover, neuroblastoma may change into ganglioneuromas[10]. Ganglioneuromas more commonly develop in females[10]. Present case also developed in a female child. Ganglioneuroma may also originate in parapharyngeal space[11]. It is believed that ganglioneuroma originates as a result of maturation of neuroblasts into ganglion cells[12]. Rarely, ganglioneuroma may contain leydig cells and may also produce virilization[13]. Ganglioneuroma may occur spontaneously or during treatment of neuroblastoma with radiation and/or chemotherapy. Thus, ganglioneuroma appears to be the benign example of neuroblastoma[14]. Cytoplasm of ganglion cells may contain neurosecretory granules and uncommonly form the synapses[15]. Ganglioneuromas (GN) are tumours of sympathetic nervous system arising from neural crest cells[16]. GN consist of proliferated ganglion cells embedded in schwannian stroma. Pre-sacral retrorectal location of ganglioneuroma is very rare[17]; only 20 cases of ganglioneuromas at this anatomical site have been reported. First case of peripheral nervous system ganglioneuroma was described by Leeson and Hite in the year 1870[18]. GN commonly develop in older children, aged >7 years[19]. However, the current case was aged 5 years. Few ganglioneuromas may be hormone-secreting; the tumour may secrete catecholamines which may produce flushing and hypertension. Moreover, ganglioneuromas may produce local burden and may produce mass effect to cause pain or paresis. Ganglioneuroma is a well-differentiated tumor constituted by mature ganglion cells. Some patients may develop lower abdominal pain and constipation/diarrhoea due to local compression of rectum as well as lumbar and sacral plexus[16]. In the present case, diarrhoea was not seen. Moreover, the current patient did not have neurological or neuroendocrine secretory symptoms. In addition, present case was not associated with any of the hereditary syndromes, eg. MEN 2, Cowden syndrome, familial polyposis and tuberous sclerosis. Additionally, micro calcifications and focal B-cell and plasma cell infiltration may occur in ganglioneuromas. In the current case also perivascular cuffing and diffuse lymphocytic infiltration were seen. Rarely, complications may develop following its proximately with major vessels[20].

Conclusion

Ganglioneuroma appears to be a benign schwann cell-dominant tumour comprising of mature ganglion cells, schwann cells and stroma. Clinical symptoms develop when the tumour compresses the surrounding tissues. Rarely, the tumour may be hormone-producing. Surgery is the treatment of choice and may result in complete cure. Large number of patients with ganglioneuroma may be cured after excision of tumour mass.

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