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Case Report

Presacral ganglioneuroma in an adult with 6-year follow-up without surgical treatment[☆]

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ABSTRACT

Ganglioneuroma is a rare tumour originating from neural crest cells, occurring mainly within children older than 7 years. It can be localised in pelvic; however, this localisation is extremely rare. This paper presents the case of a 39-year-old woman, at whom the pelvic localisation of the lesion and the unspecific symptoms associated with the digestive and genital tract impeded the recognition of the actual disease. The immensely slow growth of the tumour, combined with gradual fading of the symptoms, indicated its benign character. Only the CT-controlled biopsy enabled the recognition of the ganglioneuroma. Taking under consideration the histopathologic result and the cease of the symptoms, we decided to leave the patient under observation. After 6 years of observation, no progression signs have been recorded.

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Introduction

Ganglioneuroma is a rare tumour originating from neural crest cells. It is the most mature form of the neuroblastic tumours and it accounts for 0.1% to 0.5% of all tumours of the nervous system [1].

Ganglioneuroma is predominately detected in children older than the age of 7 [2]. The most common lesion

locations are: chest, in particular posterior mediastinum (41.5%), extraperitoneal space (37.5%), adrenal glands (21%) [1–3]. Chest and extraperitoneal ganglioneuroma is most commonly recognised in older children, while the adrenal location is characteristic of patients between 30 and 50 years of age [1–6]. The pelvic localisation is extremely rare. There have been about a dozen reports of the pelvic tumour localisation by 2008 and by less than 20 by 2016 [7–10].

[☆] In Poland the consent for treatment is also the consent for data publication in this type of papers

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The tumour usually appears in areas with large vessels in the course of the trunk of the sympathetic system [1,6].

Some ganglioneuromas can also be hormonally active, that is, secrete catecholamines, that can affect the course of disease [3]. Despite its usual benign course, ganglioneuroma can suppress other structures while growing, causing pain and/or irreversible paresis.

The treatment of choice is surgical excision. After surgery, the tumour may relapse [12].

Case report

A 39-year-old woman reported to the emergency room (2014, May) complaining about a certain paroxysmal pain located in the hypogastric region and diarrhoea occurring in turns with bowel obstruction. The pain was not ceased after the intake of the common over-the-counter analgesics. The abdominal ultrasonography showed no abnormalities. She has been admitted to the hospital and subjected to diagnostic tests. The morphologic and biochemical parameters were normal. Gastroscopy and colonoscopy were performed. A flat, 6-mm polyp was found in the transverse colon and resected. Result of histopathologic examination showed benign adenoma. After receiving the conservative treatment that has improved the patient's state, the woman has been sent home.

In September 2014, a benign cyst in right ovary has been diagnosed and removed by laparoscopy. A recurring pain in the hypogastric area was detected; therefore, it was decided to perform the magnetic resonance imaging (MRI). The examination showed the presence of longitudinal, solid lesion with dimensions: 106 × 26 × 33 mm in the presacral area, including the aortal bifurcation, and running down, unilaterally along the left iliac common vessels. The light of the vessels was not narrowed. The MRI protocol used included the turbo spin-echo T2-weighted images, fat-suppressed T2-weighted, Turbo Inversion Recovery (T2-TIRM), Diffusion Weighted Echo-Planar Imaging (DW EPI), pre- and postcontrast dynamic 3D T1 Gradient Echo (GRE) in transverse orientation.

The lesion had a hypointense signal in T1- and T2-weighted imaging sequences, and it was respectively homogeneous and heterogenous (Fig. 1). There was no diffusion restriction in Diffusion Weighted Imaging (DWI) sequences (Figs. 2 and 3).

In STIR sequence, the tumour was heterogenous and it had hyperintense signal. In a postcontrast dynamic imaging, the lesion had a moderate and heterogenous enhancement (Fig. 4). While performing the differential diagnosis sclerosing mesenteritis, demoid type tumour, lymphoma and carcinoid have been taken into account.

Due to the unclear outcome of the MRI, computed tomography (CT) has been performed. The examination outcome showed the lesion in normal density (about 40 HU) with minor, heterogenous enhancement in the postcontrast phase (at most to 60 HU). There were no calcifications and decay foci (Fig. 5).

Taking into consideration the moderate clinical manifestations and laboratory test results and imaging methods, the patient has been put under observation.



Fig. 1 – Initial study (November 2014). Sagittal T2-weighted fast-recovery fast spin echo MR—homogenous and heterogenous mass.

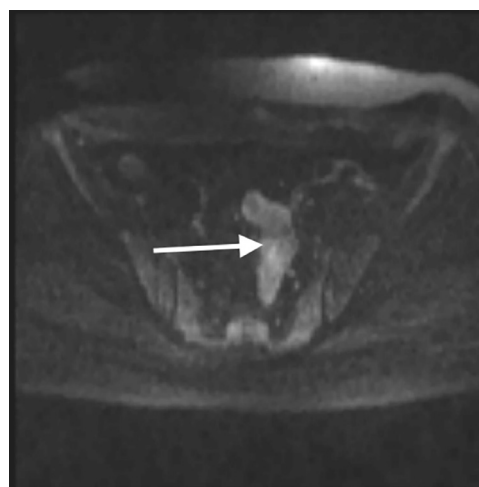


Fig. 2 – Initial study (November 2014). Diffusion-weighted MR image (b = 800). Hyperintense mass without restricted diffusion.

During 2 years of the observation, the MRI showed a slow increase in one of the lesion's parameters and the infiltration embracing the left parametrium. The laboratory results showed minor thrombocytosis (393 G/l), a slightly elevated CA-19.9 level (35.58 IU/mL), and normal levels of the CEA, CA-125 and CA-15.3, as well as a normal serum concentration of catecholamines and their metabolites. Therefore, it was decided to perform a CT-guided core biopsy (Figs. 6 and 7). Histopathology report showed a neuronal tumour in type of ganglioneuroma.

The previous pain in the hypogastric area has subsided. Due to the benign character of the tumour and the lack of clinical symptoms no surgical treatment has been performed. The patient is currently followed up. The regular MRI and CT check-ups did not show any progression (Figs. 8 and 9). In addition, the pelvic pain symptoms subsided.

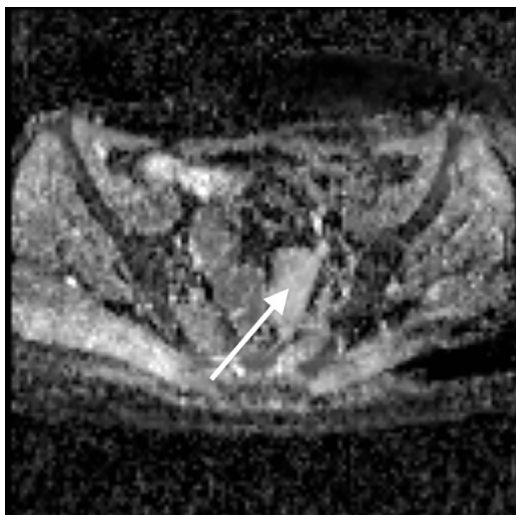


Fig. 3 – Initial study (November 2014). ADC map with the region of interest show no restricted diffusion.

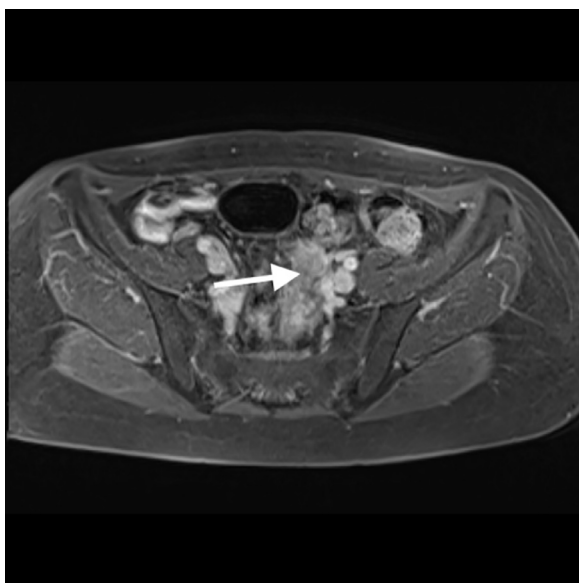


Fig. 4 – Initial study (November 2014). Axial contrast enhancement T1-weighted MR with fat-sat show mass with moderate and heterogenous enhancement.

Discussion

Ganglioneuroma is a rare tumour, originating from neural crest cells. It belongs to the neuroblastic malignancies, such as neuroblastoma and ganglioneuroblastoma, and is the least malignant one. It also has the richest schwannian stroma, which surrounds single mature ganglion cells. According to Shimada, ganglioneuroma develops as a result of maturation of neuroblastoma [1]. There is no evidence that all ganglioneuromas are formed this way, but more research seems to confirm this way of its development [12]. The location of ganglioneuroma tumours described in our work is extremely rare.

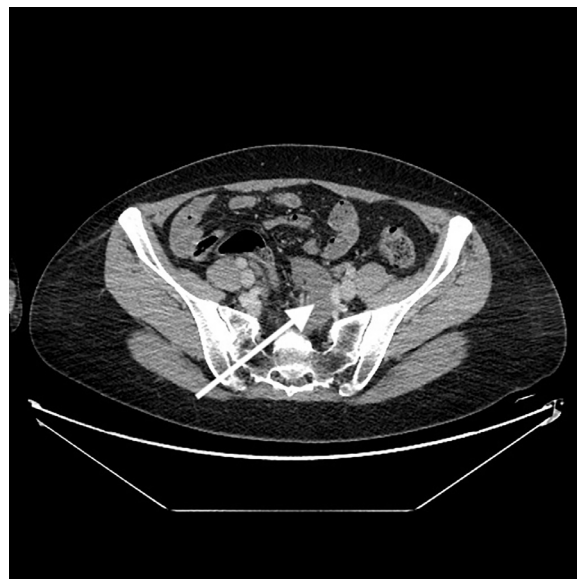


Fig. 5 – Axial contrast enhancement CT image shows a bulky mass with moderate enhancement with no calcifications and decay foci (December 2014).



Fig. 6 – Axial CT-guided core needle biopsy (September 2016).

By 2016, less than 20 cases in the pelvic location were described [10].

In this case, laboratory tests did not deviate from the normal values. Due to the localisation, the ultrasound check-up had a limited diagnostic value. It was necessary to extend the diagnosis to the MRI and CT. The check-up result of just one method may not be reliable and conclusive, but undertaking both CT and MRI, we can differentiate the tissues of the ganglioneuroma with a large component of ganglion and Schwann's cells and the connective tissue stroma. Same as in the other authors' works, the density was higher than 30 HU in CT scans and the MRI has showed the heterogenous, hypointense signal in T2-weighted imaging [6,15]. Also, in post-contrast phases, the enhancement was much more distinct in the MRI than in the CT [16,17]. The lesion was located along the blood vessels, but did not surround them and did not cause any narrowing of their light. These characteristics may have

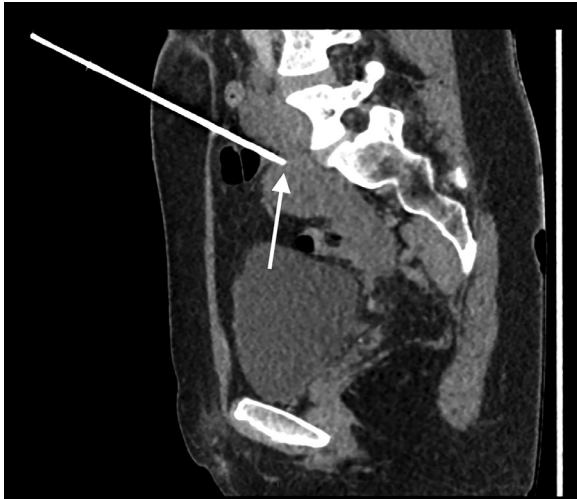


Fig. 7 – Sagittal CT-guided core needle biopsy (September 2016).

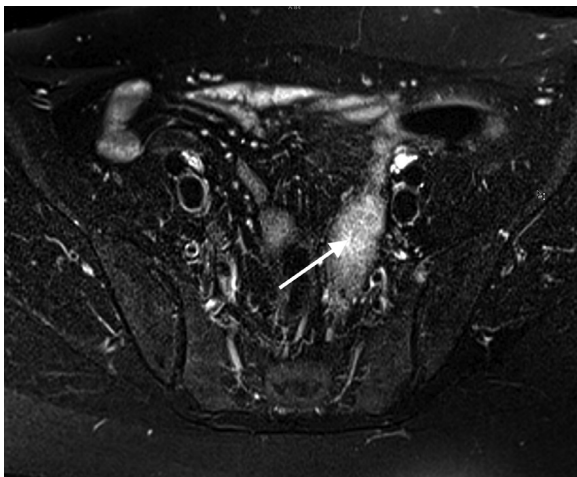


Fig. 8 – Last study (December 2019). Axial T2-weighted MR STIR, hyperintense mass-stable disease.

indicated the benign nature of the tumour, just as in children's cases [15].

Due to the unusual location of the tumour, difficult diagnosis and the slow, but steady increase in tumour's size in the MRI, it has been decided to perform a coarse-headed biopsy. Nishihira et al. suggest that the slow growth of the tumour may be result of stromal Schwann's cells' proliferation and not the hyperplasia of malignant tissue [11,13]. This could explain the, observed in subsequent MRI, slight increase in the size of the tumour, without any evidence of disease progression – as the tumour's growth is not greater than 25%.

The method of choice in the treatment of ganglioneuroma is a surgery with total or subtotal resection of the tumour, having a similar prognosis to the incomplete resection with less than 2 cm residual disease [13,14]. Adjuvant chemotherapy does not affect the progression-free survival or overall survival in any way. The benefits of chemotherapy occur only in cases that include immature cells and only within the tumours that

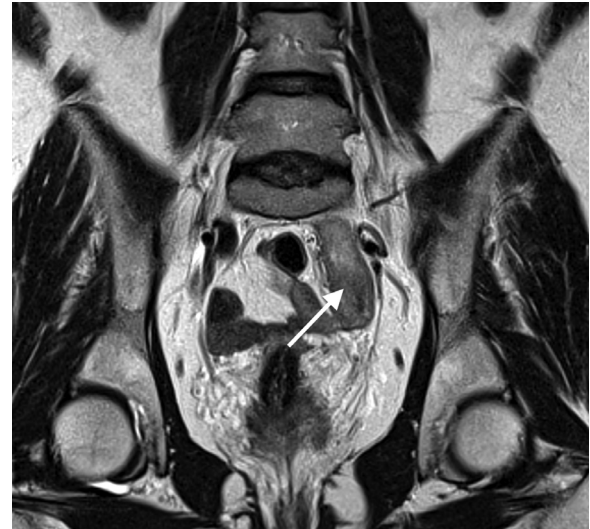


Fig. 9 – Last study (December 2019). Coronal T2-weighted MR, heterogenous mass-stable disease.

have not been resected completely [12]. If the prognosis shows that the excision would not cause any clinical improvement and the surgery is more risky than the disease's progression, the observation appears to be the only right solution.

It is not unusual for the ganglioneuromas to relapse in the postexcision [12].

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