Case Report

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Pre-sacral glomangioma: a rare localization of glomus tumors: case report

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Introduction and importance: Deep-located glomangiomas are rarely reported. Because of their scarcity, treatment strategy is hard to establish. Herein, the authors report the first case to our knowledge of pre-sacral glomangioma.

Case presentation: A 34-year-old female patient, with no previous medical history, consulted for 2-month-old pelvic abdominal pain, vomiting and delayed menstruation. Her physical and biological parameters were with no abnormalities. MRI of the pelvis demonstrated a 14 cm mixed heterogeneous pre-sacral lesion pushing the rectum anteriorly. She was operated on via a laparoscopic approach. Division of Douglas' pouch and pelvic peritoneum laterally to the bladder showcased a cystic lesion of 13×8 cm occupying the pelvis while deviating the rectum anteriorly. Its content was aspirated and left membrane was extracted in a sac. The postoperative course was uneventful.

Discussion: Pre-sacral masses are hard to treat because of their large heterogeneity. Surgical resection should be tempted to retrieve the definitive histological diagnosis and relieve the patient. However, the surgical route is controversial as each approach has its advantages. Thus, the surgical route should take into consideration the lesion's size, height and surrounding contacts, the patient's functional state and surgeon's expertise.

Conclusion: Pre-sacral glomangiomas carries a low malignant pattern but should be resected to offer histological diagnosis. The surgical route remains at the surgeon's decision, with the main objective to totally resect the encountered lesion without causing functional and sexual complications or harm to surrounding viscera.

Keywords: case report, glmangioma, Kraske, pre-sacral, retro-rectal, tumor

Introduction

Literature is filled with digital glomangiomas. Reports of extra-digital ones are confined to peripherical segments. Deep-located cases are rarely observed. Herein, we report a glomangioma localized in the pre-sacral space. We aim to shed lights regarding this affection, with a special emphasis in its diagnosis and treatment. Surgical resection is the standard therapeutic course[1], as it precludes tha hasards of cancerous lesion. In fact, the invasive potential of pre-sacral tumors is variable, but often benign in nature (92%)[2]. But, the surgical route is controversial as each approach harbors its advantages. Bearing in mind each's procedure risks with a good incorporation of patients' comorbidities and risks, will render the best results. This work has been reported in line with the SCARE 2023 criteria[3].

HIGHLIGHTS

- Deep-located glomangiomas are rarely reported.
- This is the first case of pre-sacral glomangioma.
- Preoperative diagnosis is delicate, as clinical and paraclinical features lack specificity.
- Surgery is the backbone of its treatment.
- Surgical route is controversial and should be tailored to patient's and tumor's characteristics.

Case report

A 34-year-old female patient, with no previous medical history, sought medical care for 2-month-old pelvic abdominal pain and vomiting. The pain was mild, dull and worsend when sitting. She reported delayed menstruation. She had no known allergies neither hereditary disease. She denied smoking or alcohol intake. Upon examination, her blood pressure was 100/60 mmHg, she had 64 pulse-per-min.

On physical examination, she was afebrile and no mass was perceived.

Her biological parameters were within normal range with hemoglobin level at 12.2 g/dl.

MRI of the pelvis demonstrated a mixed heterogeneous pre-sacral lesion (tissular and cystic). It was well defined, measuring 90×67×140 mm and enhanced after Gadolinium intake. A left

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latero-vesical extension was noted and the mass pushed the rectum anteriorly (Fig. 1).

Because of the fear of tumoral progression and its symptomatic feature, resection was deemed necessary, after discussion with the patient. Thus, she was operated on, in our general surgery department, via a laparoscopic approach. Division of Douglas' pouch and pelvic peritoneum laterally to the bladder showcased a cystic lesion of 13×8 cm occupying the pelvis while deviating the rectum anteriorly. Its content was aspirated and the cystic membrane was extracted in a hermetic sac. The postoperative course was uneventful and was discharged on the 3rd postoperative day. We are now 10 years after the index she is recurrence-free on repeated and regular follow-up.

Histological examination was consistent with a glomangioma without cancerous features. It showed heterogeneous pattern associating dense cellular sections made up of monomorphic round glomus cells within a grafted and vascularized stroma, with myxoid zones of low cellularity (Fig. 2). Glomus cells have regular nuclei with fine chromatin and eosinophilic cytoplasm (Fig. 3).

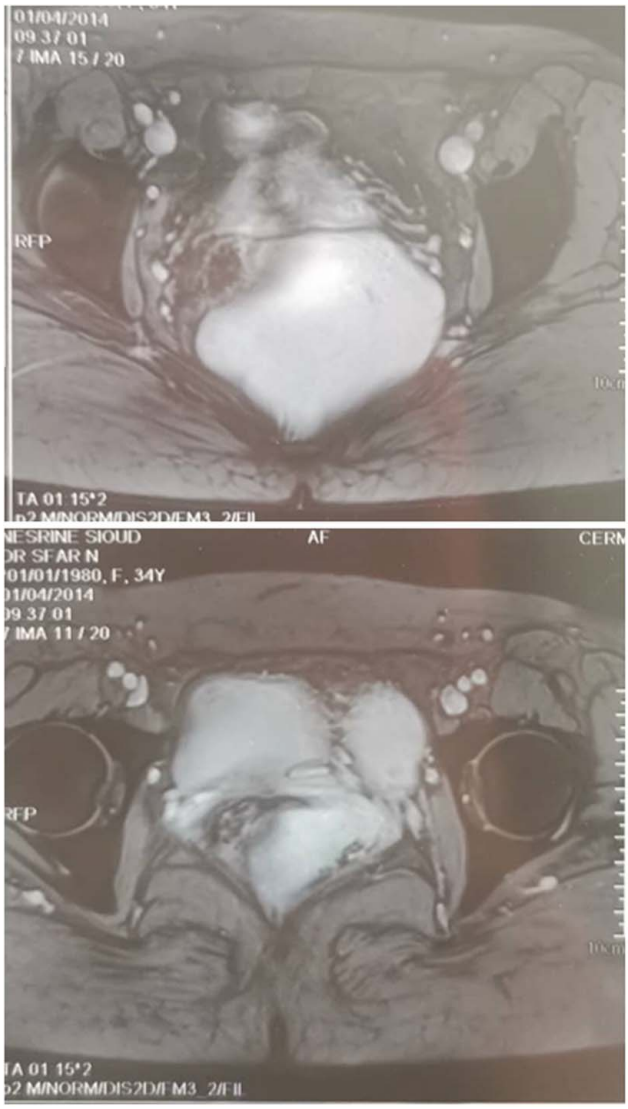

Figure 1. MRI images demonstrating a well-defined mixed heterogeneous presacral lesion (tissular and cystic) measuring 90×67×140 mm and enhanced after Gadolinium intake. A left latero-vesical extension was noted and the mass pushed the rectum anteriorly.

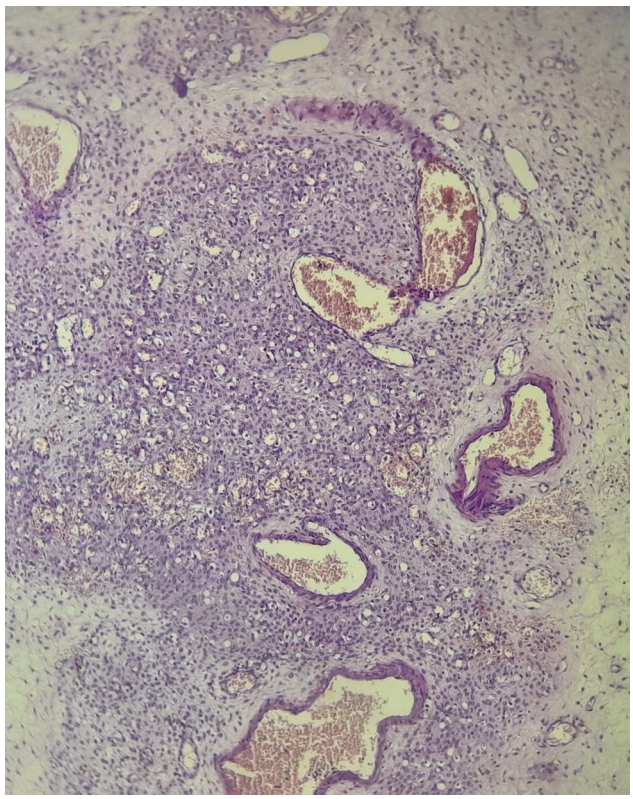

Figure 2. Microscopic picture (hematoxylin eosin ×10) exhibiting round-cell tumor proliferation in the periphery of blood vessels with dilated lumen.

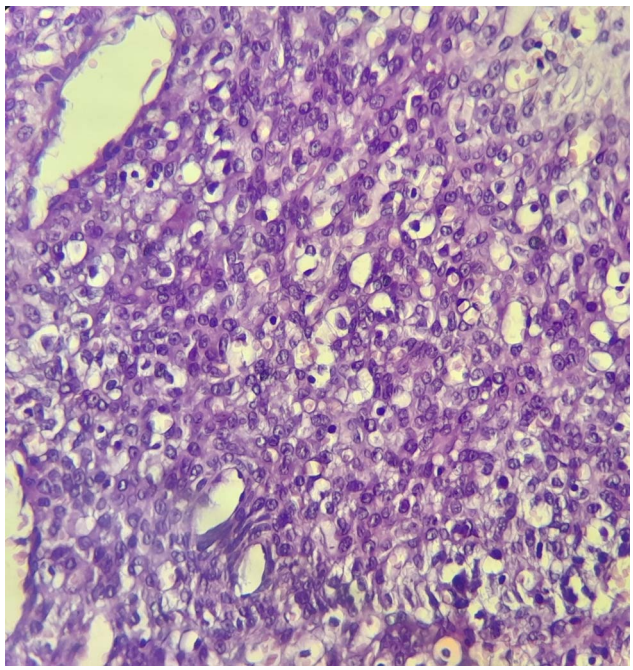

Figure 3. Microscopic picture (hematoxylin eosin ×40) showing rounded monomorphic glomus cells with eosinophilic cytoplasm and regular nuclei.

Discussion

Although glomus tumors are thought to be related to glomus bodies, these entities have been observed in extra-cutaneous locations that are not known to contain glomus cells[4]. They occur theoretically everywhere[5] but are predominantly cuta neous with acromelic propensity[6]. One possible explanation for this finding is that these tumors may arise from peri-vascular smooth muscle cells that can differentiate into glomoid cell types[4].

The main histological type of pre-sacral tumors is epidermoid cyst (34%), and the rest of the tumors are mainly benign in nature[7]. Our case illustrates, to our knowledge, the first pre-sacral glomangioma.

Even if malignant potential is not doubtful, surgery is required whenever it causes functional or clinical repercussions, or to enable definitive histological diagnosis, or because of the risk of malignant transformation into adenocarcinomas and neu-roendocrine tumors, like it was observed in others retro-rectal cystic tumors such as tailgut cyst[8].

Their unusually deep location, as in our case, also requires an extirpative surgery because it is correlated with malignant potential[6]. According to a bi-centric study of 52 cases with malignant features, 5-year cumulative metastatic risk was significantly increased for tumors with deep location ($P = 0.004$), size more than 2 cm ($P = 0.005$), and atypical mitotic features ($P = 0.004$)[9]. In a large multicenter French series involving 270 patients from 18 specialized centers over a 20-year period, 8% of tumors removed were malignant[2]. This further underlines the importance of systematic resection of all retro-rectal lesions, regardless of their presumptive type, even though silent lesions account for 40–55% of cases[7,10]. Moreover, 68% of malignant tumors were diagnosed on the surgical specimen[2]. The perfor mance of preoperative MRI for the prediction of cystic nature, estimated by the area under the curve was 0.74 (sensitivity = 71%, specificity = 78%; $P < 0.001$), whereas it appeared insuffi cient for the prediction of malignant nature of retro-rectal tumors preoperatively (area under the curve = 0.7, sensitivity = 85%, specificity = 57%, $P = 0.06$). However, biopsy could improve diagnostic performance, with areas under the curve of 1 (sensi tivity = 100%, specificity = 100%, $P < 0.001$)[2].

Surgery should be in-toto resection, to not to leave any tumorigenic islets, which can occur in 6% of patients operated on for benign tumors versus 23% for malignant tumors[2].

Surgery is recommended, especially as it offers low post-operative morbidity (chronic pain 8%, urinary dysfunction 3%, sexual dysfunction 2.5%, anal incontinence 1%)[10]. The anterior approach is the standard for large tumors located above the third sacral piece[11]. We decided to carry out the laparoscopic approach on our patient because of the many expected benefits, in particular the high risk of intra-operative tumor perforation, which is around 42% and tops the list of intra-operative incidents when carried via Kraske's route[2]. This approach has the hypo thetical advantage of offering a better operative field day with the magnifying view of the laparoscope. Postoperative pain is not influenced by the choice of surgical route, but rather by the time since surgery[12]. No significant difference was reported in terms of overall surgical complications nor complication severity between the two groups. The laparoscopic approach was sig nificantly associated to a higher risk of rectal fistula ($n = 3/53$, 6% vs. $n = 0$, $P = 0.01$) and postoperative ileus ($n = 4/53$, 7.5%, vs. $n = 0$, $P = 0.002$), and a lower risk of wound infection ($n = 1/53$, 2% vs. $n = 21/169$, 14%, $P = 0.02$) than Kraske procedure.

However, the rate of functional postoperative complications was higher in patients operated on laparoscopically in the same cohort (urinary dysfunction and dyschezia: 3% vs. 0, $P = 0.006$ and 10% vs. 0, $P = 0.02$, respectively)[2]. This result was biased by the fact that in this study, laparoscopy was preferred in the case of large lesions, which could explain the increased intra-operative tissue attrition leading to these complications.

On histological examination, we can identify 3 tumoral sub-types based on the predominant tumor contingent[6]: solid glomus tumor (predominantly glomus cells), glomangioma (prominent vascular component) and glomangiomyoma (prominent smooth muscle components).

Numerous microscopic histological similarities were observed with other tumors. Immunohistochemical studies help to narrow down the diagnosis in case of diffuse αSMA and MSA expression (found in nearly all cases: 99% and 95%, respectively), and focal to diffuse CD34 immunostaining in 32% of cases[13]. Whereas, CD31, desmin and keratins are nearly never expressed[13]. But it appears that actin, collagen type IV and vimentin are expressed in malignant cases[9]. Malignancy should be is suggested in the case of evidence of marked nuclear atypia and atypical mitotic features[6], and thus raises strict surveillance course after surgery to detect recurrences.

In the absence of solid studies detailing their evolutive profile, no current scientific data advocate the addition of postoperative treatment or peri-operative management. As a result, further studies will be carried out to establish a therapeutic plan adapted to each presentation.

Our report is the first of its kind, thus no conclusions could be compelled. But it can gleam future reports by providing evidence-based judgments.

Conclusion

This is the first reported case of pre-sacral glomangioma. The decision to operate remains at the surgeon's discretion given the absence of factual data and the fact that it is not consensual, but the obsession must be total removal to avoid subsequent recurrence.

Ethical approval

This case report was reviewed and approved by our insitutional ethics' community.

Consent

Written informed consent was obtained from the patient for publication and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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