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Inflammatory Pseudotumor of the Urachus

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ABSTRACT

Urachal diseases often create serious diagnostic dilemmas owing to their uncommon occurrences and the diversity of their presentations. We present a rare case of inflammatory pseudotumor of the urachus. The rarity and confusing nature of this condition prompted us to submit the present information.

INTRODUCTION

Urachal diseases often create serious diagnostic dilemmas owing to their uncommon occurrences and the diversity of their presentations. This is especially true for its tumors. Malignant tumors are common among all the tumors of the urachus, of which adenocarcinoma is the most common. Benign tumors, however, are uncommon in this vestigial organ [1]. Inflammatory pseudotumor is a benign, fibromuscular tumorous growth in which there is proliferation of plasma cells, lymphocytes, and histiocytes in a benign-looking, spindle-shaped stroma (myofibroblasts). It is commonly seen in abdominal and pelvic structures. Its occurrence in the urachus has been reported only a couple of times in the literature. We present a rare case of inflammatory pseudotumor of the urachus, which was also unique for the diagnostic dilemmas it created for the treating team. The rarity and confusing nature of this condition prompted us to submit the present information.

CASE REPORT

A male child of 12 years of age presented at our surgical department with complaints of a lump in the lower abdomen for 2 months, and pain over the lower abdomen for the previous 3 days. There was no history of vomiting, alteration of bowel habit, or urinary symptoms. On abdominal examination, a single intra-abdominal lump of approximately 10 cm by 8 cm in size was found occupying the mainly hypogastric and umbilical regions, along with adjacent areas of iliac fossae, which did not move with respiration. Palpation revealed a non-tender mass, which was of normal temperature and firm-to-hard in consistency. It had a bosselated surface and ill-defined margins in its lower part. Percussion revealed dullness over the lump. The liver was not enlarged, the spleen was not palpable, and

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there was no other apparent abnormal finding.

On routine investigation, total and differential white blood-cell counts were normal, as was the routine examination of urine. Fine-needle aspiration was done and the cytology report was inconclusive. Ultrasound examination revealed a solid, multinodular mass of 63 mm by 71 mm by 63 mm in the right iliac fossa, extending up to the umbilicus. On CT scan, a large, solid, lobulated mass of 11 cm by 6 cm was seen in the pelvis (more towards the right side), which was slightly hypodense in attenuation. The mass was seen displacing bowels and indenting the bladder. It was continuous with anterior abdominal wall muscle. On IV contrast administration, the mass showed heterogeneous enhancement with sparing of the central region (dense fibrosis).

Based on these findings, an exploratory laparotomy was performed. The laparotomy revealed a large lobulated mass, which was free from adjacent bowel loops and mesenteries. Its lower part was attached to a dome of the urinary bladder in a pedunculated form (Figure 1). It was excised after securing hemostasis and was sent for histopathological examination. Postoperative recovery was uneventful.

Figure 1. Inflammatory pseudotumor on right hand and urinary bladder with Foley catheter bulb on left hand.
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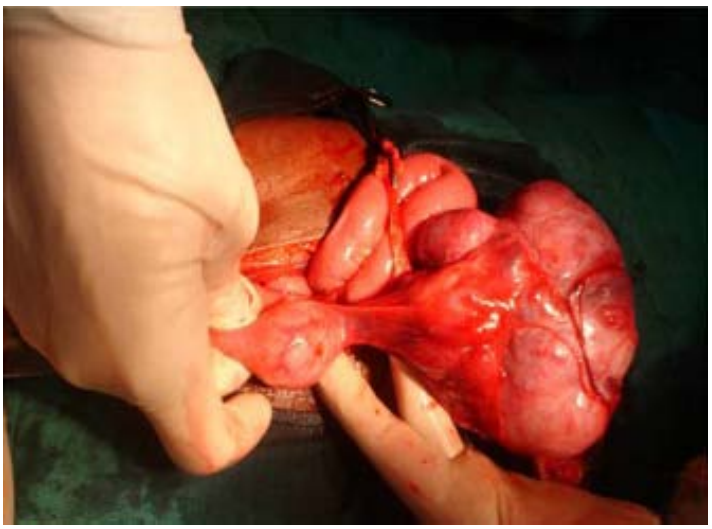

Histopathological examination of the specimen grossly revealed a greyish-white, soft-to-firm mass. On microscopic examination, the section revealed a spindle-cell tumor composed of loose fibromyxoid stroma with spindle-and-stellate-shaped cells mixed with dense lymphoplasmacytic and mononuclear infiltrate. Although the spindle cells showed mild pleomorphism, there was no mitotic activity. A focus of ischemic necrosis was also seen. The final pathological diagnosis came out to be a spindle cell tumor suggestive of inflammatory pseudotumor.

DISCUSSION

The urachus is a vestigial remnant of 2 embryonic structures, which are urogenital sinus (the precursor of urinary bladder) and allantois (the derivative of yolk sack). Normally, it is obliterated before birth, leaving a fibrous band extending from the dome of the urinary bladder to the umbilicus. Common pathologies of this structure are congenital anomalies and infections [2]. Tumors are rare findings in the urachus. The majority of tumors in the urachus are malignant (0.5% of all bladder carcinomas), adenocarcinomas constituting the majority of this group (80%) [1]. Benign tumors of the urachus are extremely uncommon. This group consists of adenomas, fibromas, fibroadenomas, fibromyomas, and hamartomas [2]. These benign tumors must be thoroughly evaluated owing to the potential confusion they create in the diagnosis of abdominal lumps as a whole [3,4,9]. Inflammatory pseudotumor of the urachus is one such tumor, which runs a benign clinical course. The most common site for this pseudotumor is the lung and mediastinum, followed by extrapulmonary sites, the list of which includes almost every organ in the body [5,6].

Despite its common predilection for abdominal and pelvic organs, including the urinary bladder, bowel, liver, and peritoneum [6,7], it has been rarely reported in the urachal remnant [8,9]. Along with the rarity of this tumor, our case is a good example of the diagnostic confusion the lumps of the urachus can create for a clinician. It resisted complete diagnosis until imaging investigations, exploratory laparotomy, and histopathology were correlated. Thus, the importance of investigations in urachal lumps is again underscored here, which was stressed previously [4,5].

Finally, regarding the treatment of inflammatory pseudotumor, 2 schools of thought are advocated. Our treatment of simple excision was due to the history, clinically as well as preoperatively, which was more in favor of a benign tumor. Simple excision has been used previously in similar cases [10]. A second school of thought also advocates that in cases where preoperative findings are inconclusive or in favor of malignancy, aggressive surgery should be the choice [9]. The best intervention frequently depends on the discretion of the

individual surgeon; however, aggressive surgery is preferred over simple excision if there is any doubt about the presence of malignancy.

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