

Angioleiomyoma in the Ischioanal Space

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We report a patient with angioleiomyoma in the ischioanal space or ischiorectal fossa. It is very unusual for this tumor to be located at this site. In fact, our review of the literature revealed that no case has been reported. Therefore, this case study is the first to report on angioleiomyoma in the ischioanal space. The definitive diagnosis for this condition is based on histological findings after extirpation of the tumor. Surgical excision is an accepted treatment for this tumor. A 47-year-old man presented with a two-month history of a progressive, palpable, painless mass in the perianal area. The ischioanal space was investigated by physical examination, computed tomography, followed by confirmation of angioleiomyoma with surgical excision and histological analyses. Even though it is a rare condition, angioleiomyoma in the ischioanal space should be included in the differential diagnosis of an ischioanal tumor, especially when the tumor is a noninflammatory primary mass.

Key words: angioleiomyoma, ischioanal space, perianal tumor

INTRODUCTION

A tumor of the ischioanal space or ischiorectal fossa is often a direct secondary extension of a primary anorectal, prostatic, pelvic, or sacral tumor.¹ Typically, patients with an ischioanal lesion present with a perineal-gluteal swelling or mass. Computed tomography (CT) and magnetic resonance imaging (MRI) can define the anatomic origin, extent, and radiologic features of a given lesion. Angioleiomyoma in the ischioanal space is extremely rare. Despite improved imaging modalities, a clarified preoperative diagnosis is difficult. The definitive diagnosis is based on histological findings after excision of the tumor.² In fact, our review of the literature revealed that angioleiomyoma in the ischioanal space has not been previously reported. Thus, we report the first case of an adult with angioleiomyoma in the ischioanal space, which was confirmed histologically after surgical excision.

CASE REPORT

A 47-year-old man presented with a solitary, painless, palpable mass in the perianal area for two months. Digital examination revealed a firm, painless mass in the left side of the perianal area measuring about 4 cm×3 cm×3 cm, and 2 cm distal to the anal verge. The patient reported no history of any medical disease. Contrast-enhanced CT scans were conducted, which revealed a well-encapsulated homogeneous, isoattenuating mass on the left side of the perianal space (Fig. 1). The patient was diagnosed with a benign primary tumor of the ischioanal space and the tumor was excised with a wide margin of tissue around its

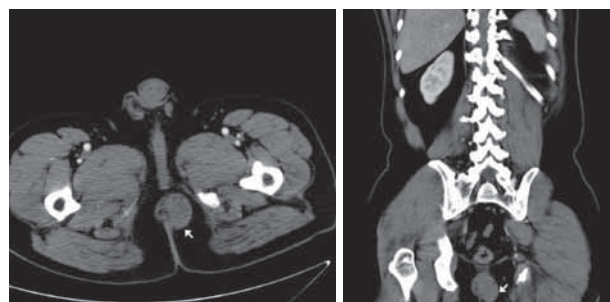


Fig. 1 The contrast-enhanced pelvic computed tomography scans revealed a well-encapsulated homogeneous, isoattenuating mass (white arrow) at the left side of perianal space.

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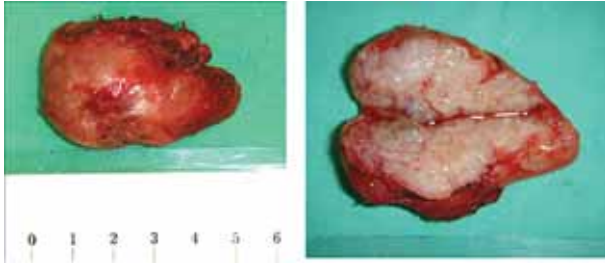


Fig. 2. In gross, the encapsulated mass was 5.0 cm×4.0 cm×3.5 cm, and the transverse section showed a firm,

perimeter. The gross anatomy of the encapsulated mass was 5 cm×4 cm×3.5 cm, and cut sections of the mass were firm, pale, fawn-colored tissue (Fig. 2). Microscopically, the mass was lined with a thin fibrous capsule consisting of bundles of plump, spindle-shaped smooth muscle fibrils with cigar-shaped nuclei and numerous thin-walled vessels. Immunohistochemically, the tumor cells were diffusely positive for desmin and smooth muscle actin. Hematopoietic progenital cell antigen (CD34) was absent, except for in the endothelial cells lining the vascular spaces. Immunostaining for CD117 (c-kit), estrogen receptor (ER), and progesterone receptor (PR) were negative. There were a few vessels with smooth muscle bundles surrounding the vessel wall. The vessels were large in number and small in size (picture CD34). This is compatible with a solid type of angioleiomyoma (Fig. 3). The postoperative course was uneventful and there was no local recurrence during a two-year follow-up period.

DISCUSSION

Angioleiomyoma, a benign tumor with vascular and smooth muscle components, rarely occurs in the ischioanal space. To our knowledge, this is the first reported patient with angioleiomyoma in the ischioanal space. Previous studies have reported that most angioleiomyomas are usually found in the four extremities (74%), and in the head and neck areas (13.1%).²⁻⁵ The clinical symptom of pain is related to the histological type of the tumor.²⁻³ Pain is experienced in 70% of patients with the solid type tumor, whereas 70% of patients with the venous or cavernous type tumor experience no pain. However, our patient had the solid type of angioleiomyoma and had no pain. We propose that ischioanal angioleiomyomas do not cause symptoms until the tumor is large.

Preoperative diagnosis of ischioanal angioleiomyoma

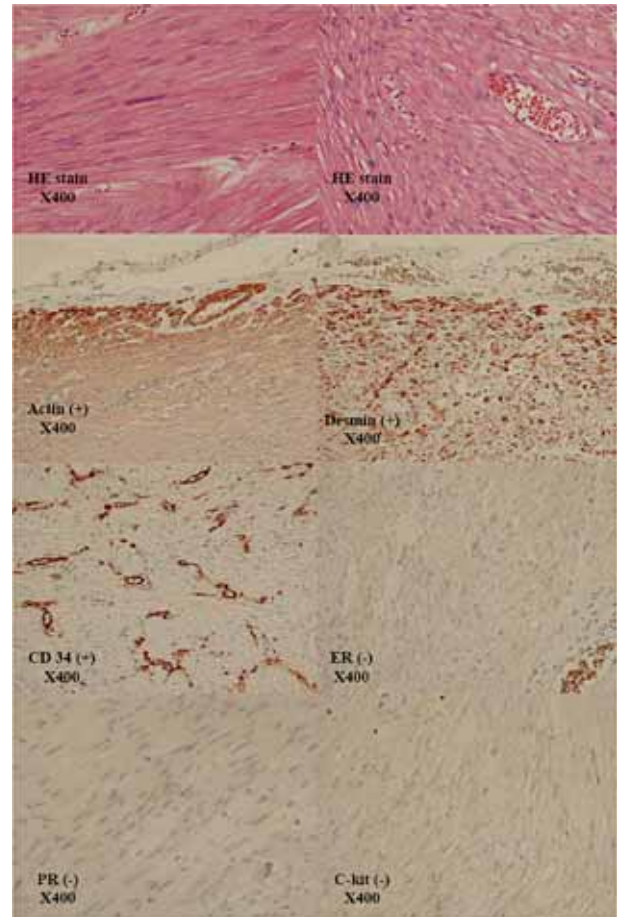


Fig. 3 Immunohistochemical stains of the angioleiomyoma were diffusely positive for desmin and smooth muscle actin (SMA). Hematopoietic progenital cell antigen (CD34) was absent, except for in the endothelial cells lining the vascular spaces. Immunostains for CD117 (c-kit), estrogen receptor (ER), and progesterone receptor (PR) were all negative. The vessels were large in number but small in size (picture CD34). This is compatible with a solid type of angioleiomyoma.

is difficult. In our patient, the contrast-enhanced CT scans showed a benign, well-encapsulated homogeneous, solid, isoattenuating mass. For this reason, MRI was not performed. Nevertheless, MRI is the modality of choice because of its excellent contrast resolution and multiplanar ability.¹ CT and MRI are useful in differentiating deep-seated angioleiomyoma from malignant or other benign tumors, such as lipoma or fibroma.⁶⁻⁷ Most angioleiomyomas can be diagnosed correctly with conventional histology using hematoxylin and eosin staining. Histochemical stains for smooth muscle cells, such as Masson's trichrome, and immunochemical stains for smooth muscle

cells, such as desmin or actin, and for the endothelium of vessels, such as factor VIII-related antigen, CD34, or CD31, can be used to differentiate between angioleiomyoma and other spindle cell-shaped tumors, such as hemangioma, angiofibroma, fibroma, and angiomyolipoma.⁸ In our patient, the differential diagnosis included aggressive angiofibroma (actin +, desmin +), gastrointestinal stromal tumor (c-kit +), fibroma and angiofibroma (myxoid stroma), and angiofibrosarcoma (ER +, PR +, female-predominant, myxoid stroma).

The definitive diagnosis was based on histological findings after extirpation of the tumor. Local excision is an accepted treatment for this condition.² In our patient, the tumor was well encapsulated without extracapsular extension. Consequently, local excision along the capsule of the tumor was adequate to remove the angioleiomyoma from the ischioanal space. The postoperative course was uneventful and there was no local recurrence during a two-year follow-up period.

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