Answer

Presacral Neoplasm

Presacral neoplasms are rare, with an incidence of 1 in 40 000 hospital admissions.[1,2] Following Uhlig and Johnson's[1] classification scheme, presacral tumors are categorized as congenital, neurogenic, or osseus. Several other types of soft tissue tumors such as lipomas or sarcomas fall into the miscellaneous category. Two-thirds of all presacral tumors are congenital, and this patient's pathology was consistent with one such congential tumor, a cystic hamartoma. Two-thirds of congenital tumors are benign, and other examples include epidermoid and dermoid cysts and anterior sacral meningoceles. Sacrococcygeal chordoma, a solid tumor, is the most common malignant congenital lesion in the presacral space.[3]

Because most presacral tumors have an indolent course, they are often discovered incidentally on periodic rectal examination or on imaging studies obtained for other reasons. The Mayo Clinic reported that almost all (97%) presacral tumors were palpable on digital rectal examination,[4] although we could not palpate the lesion in our patient until she had received general anesthesia. We suspect this was owing to her obesity and because the lesion was drained prior to our evaluation. Presacral neoplasms most commonly present with vague lower back or perineal pain that is aggravated by sitting or defecation.[4] The presence of a postanal dimple should alert the examiner to the possibility of a presacral neoplasm, especially congenital lesions like epidermoid, dermoid, or enterogenous cysts, as they frequently communicate with the skin. These congenital cysts are more common in women and are frequently misdiagnosed as anorectal abscesses of the typical cryptoglandular variety.[4] Computed tomography is the optimal imaging method to differentiate between solid and cystic masses and to identify cortical bone destruction that is associated with chordomas.[2] Magnetic resonance imaging complements computed tomographic findings by providing enhanced soft tissue resolution and identifying cord anomalies such as a meningocele.[5]

The indications for preoperative percutaneous biopsy are somewhat controversial. In general, biopsy should be reserved for solid tumors with imaging characteristics that make them suspicious for malignancy. Preoperative tissue diagnosis in some solid tumors may guide surgical margins or dictate preoperative chemoradiation therapy. Presacral lesions that are cystic rarely require biopsy. Although 10% to 38% of patients with cystic lesions contain some component of malignancy,[6] unnecessary instrumentation of these lesions may predispose them to infectious complications. If biopsy is necessary, it should never be performed transrectally.[7]

Presacral neoplasms are resected if there is a possibility of malignancy, symptoms, or (cystic lesions) infection. Furthermore, repeated procedures to drain the infections may risk incontinence. The choice of operative approach is determined by physical examination and preoperative imaging. If the examining digit can get around most of the lesion during digital examination or if the lesion is below S3 on imaging studies, they can usually be managed via a posterior approach involving a para-sacrococcygeal incision. Tumors above the S3 level may necessitate either an anterior or a combined anterior and posterior approach.[8] Some have argued that a coccygectomy must be performed for all congenital cystic lesions because the coccyx is believed to contain totipotent cells that lead to recurrence.[6,9] In this case, the patient was positioned prone and a para-sacrococcygeal incision was used. An assistant's gloved hand was inserted into the rectum to push up on the lesion and help deliver it into the operative field to facilitate dissection. The lesion was excised en bloc with its multiple cystic extensions and the overlying skin containing the dimple (Figure 2). Pathologic analysis revealed a dominant cyst with multiple satellites containing squamous, ciliated pseudostratified columnar and cuboidal epithelium. Presence of these varied epithelial tissues along with a discontinuous smooth muscle component but absence of skin adnexal structures or neural plexuses differentiates cystic hamartoma from teratomas or other congenital lesions.[10] The literature reports 43 total cases of cystic hamartoma with 17 reported cases of malignancy arising in these lesions since 1988.[10] Our patient did not have any component of malignancy in the specimen.

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