Subcutaneous Sacrococcygeal Myxopapillary Ependymoma

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Case Report

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Summary: We report a case of myxopapillary ependymoma presenting as a primary tumor of the subcutaneous tissue in the sacrococcygeal region. The mass was large, well-encapsulated, lobulated, and multiseptated, with varying signal intensity on T1- and T2-weighted MR images caused by hemorrhagic necrosis, blood degradation products, and calcification. Only a small viable portion enhanced after administration of contrast material. Multiple lobules formed from fibrous septa and dystrophic calcification also characterize this tumor.

Myxopapillary ependymomas are typically primary, intradural tumors of ependymal origin that arise from the filum terminale. In rare instances, they may arise in the sacrococcygeal region as a primary subcutaneous tumor (1–9). In this setting, the tumors may present as either a dorsal sacrococcygeal growth or a subcutaneous nodule (5, 6). We report a case of primary subcutaneous sacrococcygeal myxopapillary ependymoma seen on plain radiographs and MR images.

Case Report

A 54-year-old woman had swelling of the sacrococcygeal region that had been present for more than 40 years. She sought medical attention after noting recent growth, pain, and discomfort on sitting. Physical examination revealed a solid mobile mass in the coccygeal region and an intergluteal fold that measured 10 × 5 cm. The overlying skin was intact without ulceration.

Plain radiographs of the pelvis showed a well-defined ovoid radiopaque mass with peripheral flocculent and punctate calcifications (Fig 1A). On MR examinations, the mass was well encapsulated, lobulated, multiseptated, and located in the subcutaneous tissue of the intergluteal fold without extension to adjacent structures. MR images showed markedly heterogeneous signal intensity, suggestive of necrosis, blood degradation products, and/or hemorrhage (Fig 1B, D, and E). Only the peripheral portion of the mass enhanced irregularly after contrast administration (Fig 1C). The fibrous capsule, internal septa, and calcifications were seen as areas of low signal intensity on all sequences. The distal thecal sac, cauda equina, and conus were normal. No intraspinal tumor was evident on MR images of the lumbosacral spine. The differential diagnosis included teratoma, complicated pilonidal cyst, neurogenic tumor, and sarcoma. No preoperative tumor biopsy was done.

At surgery, the lesion was found in the subcutaneous fat tissue just below the skin and above the anal crease with no involvement of deep muscles or bony structures. It was excised without complications. No significantly enlarged vascular feeders and no sinus tract were noted. The excised mass was lobulated and hard, and somewhat hemorrhagic in appearance, and measured 9 × 7 × 5 cm. Intratumoral hemorrhage was responsible for the recent growth of the tumor. A cut surface of the gross specimen showed a well-encapsulated mass (Fig 1F). A fibrous capsule surrounded the mass, which contained a fibrous septum, and dystrophic calcifications were noted peripherally.

Histologically, most of the tumor showed ischemic necrosis except the peripheral portion. Viable tumor cells were noted at the periphery, with attenuation caused by a predominance of mucoid globules; viable cells were not found in the central portion of the tumor (Fig 1G). The tumor cells were cuboidal, and had round to oval nuclei with neither atypia nor mitotic activity, and were arranged in a single layer (Fig 1H). Immunohistochemical stain of the paraffin sections for glial fibrillary acidic protein was diffusely positive in the tumor cells (Fig 1H, inset). The final diagnosis was primary myxopapillary ependymoma.

Discussion

Ependymomas are glial tumors primarily of the brain and spinal cord, but on rare occasions may be found outside the CNS. This unusual presentation occurs in four general situations: 1) from metastases or direct extension of a primary tumor of the CNS, seen after surgical excision (9); 2) from direct extension to the soft tissue of the sacrococcygeal area from a primary ependymoma of the lower spinal cord, cauda equina, or filum terminale (10); 3) from a primary presacral, pelvic, or abdominal tumor (11); and 4) from a primary tumor of the skin and subcutaneous tissue of the sacrococcygeal area without demonstrable connection to the spinal cord or filum (2, 9).

Myxopapillary ependymomas occur predominantly in the cauda equina or conus medullaris and rarely in the pre- or postsacral region. Since the first report of an extradural myxopapillary ependymoma in 1902 (7), over 50 cases have been reported in the posterior sacral or subcutaneous region (1–8). Primary subcutaneous sacrococcygeal myxopapillary ependymomas are believed to arise from the coccygeal medullary vestige or subcutaneous ependymal rests (6, 9). The coccygeal medullary vestige, located in the caudal portion of the

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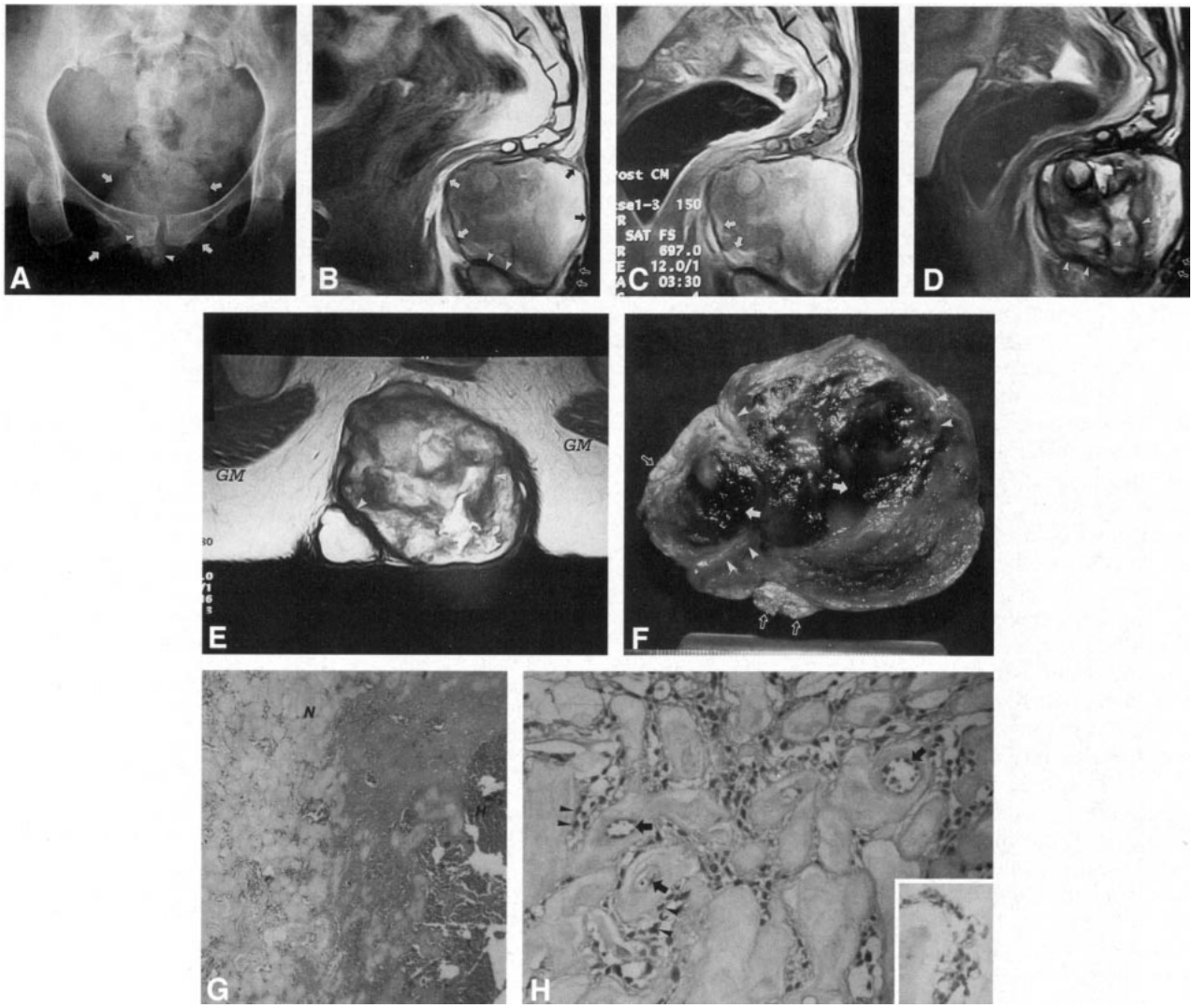

FIG 1. 54-year-old woman with subcutaneous sacrococcygeal myxopapillary ependymoma.

A, Plain radiograph of the pelvis shows a well-defined ovoid radiopaque mass (*arrows*) with flocculent and punctate calcifications (*arrowheads*).

B, Sagittal turbo spin-echo T1-weighted (600/12/4 [TR/TE/excitations]) MR image shows a well-defined, lobulated, partially septated mass in the subcutaneous tissue of the sacrococcygeal region that is heterogeneous in signal intensity. Fibrous capsule (*solid arrows*) and septa (*arrowheads*) are of low signal intensity. There are calcifications along the posterior portion (*open arrows*).

C, Sagittal contrast-enhanced fat-suppressed turbo spin-echo (500/15/3) MR image shows heterogeneous enhancement at the periphery (*arrows*). The enhancing areas correspond to the areas of high signal intensity on T1-weighted images.

D and *E,* Sagittal (*D*) and axial (*E*) turbo spin-echo T2-weighted (4000/112/3) MR images show markedly heterogeneous signal intensity, suggestive of necrosis, blood degradation products, and/or hemorrhage. Mass is divided into lobules by septa (*arrowheads*). Note calcifications in posterior portion (*open arrows, D*). *Gm* indicates gluteus maximus muscle.

F, Cut surface of the gross specimen shows a well-encapsulated solid mass that is partially divided into lobules by septa (*arrowheads*). Note hemorrhage (*solid arrows*) and calcifications (*open arrows*).

G, The viable tumor cells are noted at periphery, and become attenuated by a predominance of mucoid globules, which were not found in central portion. Hemorrhage (*H*) and necrosis (*N*) are noted in center (hematoxylin and eosin, original magnification × 40).

H, Photomicrograph shows collars of cuboidal cells (*arrowheads*) surrounding a neck of mucin with a centrally located blood vessel (*arrows*), characteristic findings of myxopapillary ependymoma (hematoxylin and eosin, original magnification × 200). *Inset:* Immunohistochemical stain for glial fibrillary acidic protein shows diffuse positivity of the tumor cells (original magnification × 200).

neural tube, is a small cavity lined by ependyma (9). Its subcutaneous site is often marked by a dimple on the skin surface. Myxopapillary ependymomas are expansile or infiltrative, with alteration or obliteration of preexisting normal structures, whereas the rests persist as small, circumscribed nodules (6).

Helwig and Stern (2) described the gross pathologic appearance of 32 cases of primary subcutaneous sacrococcygeal myxopapillary ependymomas. The mean size of these tumor was 4 cm, with a range from 1.7 to 12 cm. The tumors were generally ovoid, circumscribed, or encapsulated, and firm or rubbery in texture. A few tumors were described as soft. The cut surface usually appeared lobulated and gray-white. Other features occasionally noted included a moist appearance, hemorrhagic areas, yellow foci, cysts, and mucoid areas.

The tumors, either entirely or at least partially, had a papillary architecture on microscopy.

Few case reports have described the imaging characteristics of primary subcutaneous sacrococcygeal myxopapillary ependymomas. In one report, the mass was well defined, slightly lobulated, and isointense with muscle on T1-weighted images, with heterogeneous enhancement (8). In cases of intrasacral, intradural, myxopapillary ependymomas, the lesions were of low signal intensity on T1-weighted images and heterogeneously hyperintense on T2-weighted images, with heterogeneous enhancement after contrast administration (12, 13). The heterogeneity on T2-weighted images and on contrast-enhanced T1-weighted images may relate to fibrous tissue or sclerosis of the perivascular mucinous stroma, hemorrhage, or necrosis (13). The lesion in our patient showed heterogeneous peripheral hyperintensity, central hypointensity on T1-weighted images, heterogeneous enhancement after contrast administration, and multiple foci of low and intermediate to high signal intensity on T2-weighted images. This heterogeneity may relate to hemorrhage, blood degradation products, necrosis, or calcifications. The contrast-enhancing portion corresponded to microscopically viable tumor and the low-signal-intensity portion on T1-weighted images corresponded to necrosis. Multiple internal septa and dystrophic calcifications were also present, findings that have not previously been described in subcutaneous sacrococcygeal myxopapillary ependymomas.

Subcutaneous sacrococcygeal myxopapillary ependymomas grow slowly and therefore are often large at the time of presentation; the mean age at presentation is 17 years (range, 10 months to 47 years) (2). The differential diagnosis includes sacrococcygeal teratoma, pilonidal cyst, and neurogenic tumor. Sacrococcygeal teratomas are either cystic and solid or predominantly cystic; they are rarely solid (14). Over 50% have calcification or ossification. Sacrococcygeal myxopapillary ependymoma may be mistaken for a solid teratoma; however, most sacrococcygeal teratomas are discovered in the newborn period and imaging studies help indicate the diagnosis, especially if fat is present in the lesion (14).

Conclusion

Despite the rarity of subcutaneous sacrococcygeal myxopapillary ependymoma and the nonspecificity of imaging findings, this tumor should be considered in the differential diagnosis of a sacrococcygeal mass.

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