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Case 4366
Sacrococcygeal Teratoma with malignant transformation

Gonçalves M*, Cunha TM**

* Hospital Central do Funchal, Radiology. 9004-514 Funchal, PORTUGAL.
** Instituto Português De Oncologia de Francisco Gentil de Lisboa (IPOFGCROL), Radiology. 1099-023 Lisboa, PORTUGAL.
Hospital Central do Funchal

Section: Genital (Female) Imaging
Published: 2005, Dec. 30
Patient: 69 year(s), female

Clinical Summary

Female with a history of surgical excision of a pelvic tumor thirteen years before presented with constipation and obstruction to urinary flow.

Clinical History and Imaging Procedures

The patient referred pelvic pain associated with constipation and urinary obstruction. On rectal examination a mass was detected behind the rectum, apparently without invasion of its walls or other pelvic organs. Thirteen years before she had similar symptoms and was submitted to surgery (in another institution) for a pelvic tumor which she didn't know its histological type or precise location. A contrast-enhanced CT examination detected a cystic lesion with a parietal enhancing solid nodule in presacral location (Fig. 1). Pelvic MRI revealed a 11cm high protein content cystic lesion with a 5 cm solid nodule in its walls containing a focus of fat. The nodule enhanced with paramagnetic contrast. These MRI features were compatible with presacral teratoma with signs of malignant transformation. The lesion is adherent to the coccyx and gluteus minimus muscle, compresses the rectum, anus and levator ani muscle on the right, anteriorly displacing the vagina and uterus and extending inferiorly into the right ischiorectal fossa (Fig. 2). Surgery consisted in tumor and coccyx resection through a transverse perineal incision. The lesion was adherent to the rectum and coccyx. Within its capsule a necrotic fluid and a solid nodule were detected (Fig. 3). Histology revealed a sacrococcygeal teratoma in which the

epitelial component was adenocarcinoma (Figs. 4 and 5). Following surgery the patient was treated with radiotherapy (150 Gy). At present (4 years after surgery) the patient has no evidence of recurrence.

Discussion

Teratomas, also called dermoid cyst, are congenital tumors comprising elements derived of more than one of the germ layers: endoderm, mesoderm and ectoderm[1-6]. They originate from pluripotential germ cells, normally located in the ovaries and testis or abnormally located in the midline, as result of sequestered rests during migration of embryonic germ cells from yolk sac to gonads[1]. Teratomas result from proliferation of tissue foreign to their anatomic site instead of metaplasia. Histopathologically teratomas can be classified in[1]: a) Mature (benign): well differentiated, predominantly cystic; b) Immature (benign or malignant): undifferentiated, predominantly solid; c) Malignant (germ cell origin): choriocarcinoma, germinoma, yolk sac tumor; d) With malignant transformation (non germ cells elements): carcinoma, sarcoma. Most common locations are: gonads, anterior mediastinum, retroperitoneum, sacrococcygeal, neck and intracranial[3,5,6]. Sacrococcygeal teratomas (SCT) are the most common teratomas in newborns (1/35 000?40 000 births), and 3?4 times more frequent in females, being rare in adults[4,5,6,7]. They are classified in four types according to the amount of lesion present internally or externally: type I exclusively external (90% of SCT in newborns/children are externally visible) and type IV exclusively internal (adults)[4,7]. 60% of SCT are mature and 50-70% are detected in the first days of life. The incidence of malignant SCT in infants is 10-50%. In adults are usually benign[4,7]. Tendency towards malignancy occurs with increasing age, intrapelvic component and the amount of solid tissue[4]. SCT may be asymptomatic, cause constipation, obstruction to urinary flow, low back pain, lower extremity paresis/paresthesias, or dystocia[1,4]. The differential diagnosis of adults' SCT include anterior meningocele, rectal or anal duplication cyst, anal gland cyst, pilonidal cyst, chordoma, neurofibroma, fibrosarcoma and giant cell tumor[1,4]. On physical examination a palpable mass may be detected, compressing pelvic organs. In malignant germ cell teratoma fetal oncogenes (alpha-fetoprotein, carcinoembryonic antigen and human chorionic gonadotropin) may increase in serum[4]. Plain films may demonstrate structural abnormalities in coccyx and sacrum or calcifications[6]. Ultrasound demonstrates a tumor, with cystic and solid components (Rokitansky nodule)[3]. Fat and calcification, may be difficult to identify[5]. CT and MRI identify fat and calcium. A fat-fluid level and calcification are highly suggestive. Teeth may exist. Enhancement of solid component is a criterion of malignancy[5]. Distinguishing mature from immature teratomas is very difficult[3]. Treatment consists of surgical excision with coccygectomy (coccyx may contain totipotential cells and therefore an increased risk of recurrence)[4,6]. Recurrences (7,5-22% to 37% after total resection with or without coccygectomy, respectively) occur because of residual tumor or spillage of cyst content[4]. In our patient the previous surgery possibly was a resection of a mature teratoma, without coccygectomy. This second tumor probably represented a recurrence, which have a higher incidence of malignancy than primary tumors[4]. Additional chemotherapy and/or radiotherapy should be given in malignant SCT, yet, teratomas with malignant transformation are unresponsive to chemotherapy[6]. Close follow-up consists of complete physical examination, with

complementary CT or MRI. Serum markers may be of interest in those patients with elevated markers preoperatively[4].

Final Diagnosis

Sacrococcygeal teratoma with malignant transformation.

Figures

Figure 1 Fig. 1 - Contrast enhanced pelvic CT.

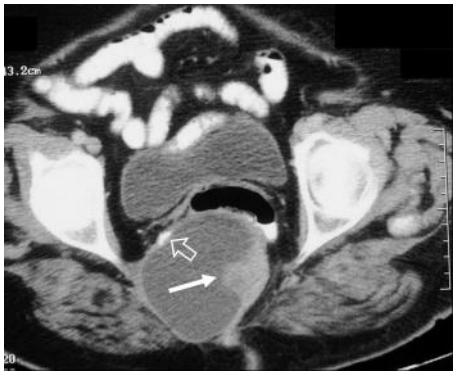

Fig. 1 - Contrast enhanced pelvic CT. A large cystic lesion with an enhancing solid nodule (arrow), has presacral location, displacing the rectum anteriorly, without signs of rectal invasion. Also a small parietal nodule containing fat and a calcification can be detected (open arrow).

Figure 2 Fig. 2 - Pelvic MRI.

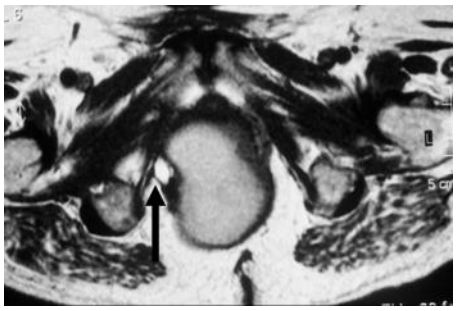

Fig. 2a - Axial T1-weighted image. A thin walled high-signal mass, anterior to the sacrum, is displacing the rectum and the vagina anteriorly. A discrete high-signal intensity lamella, detected at the right periphery of the cyst (arrow), corresponds to pure fat content, as it is completely suppressed on a fat-suppressed sequence (2b).

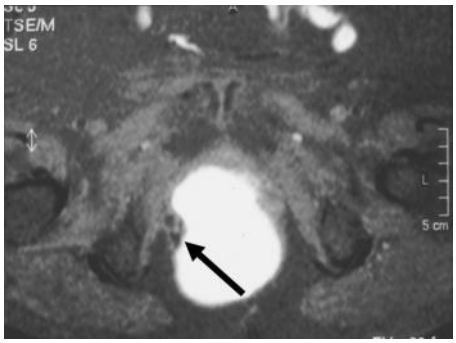

Fig. 2b - Axial fat-suppressed T1-weighted image. The high-signal intensity focus on T1-weighted image is completely suppressed (arrow).

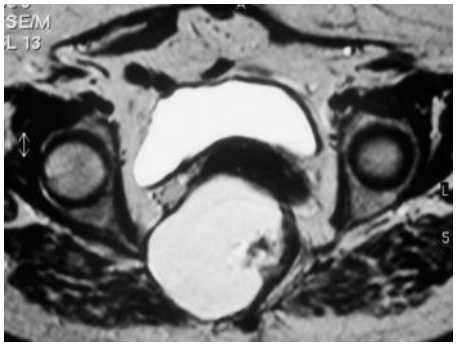

Fig. 2c - Axial T2-weighted image. The lesion has homogeneous high-signal intensity probably corresponding to an high protein content cystic mass, displacing the uterine cervix anteriorly.

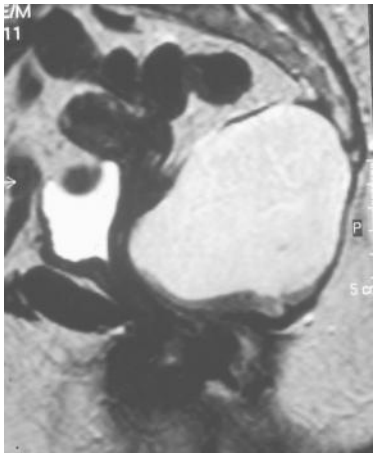

Fig. 2d - Sagittal T2-weighted image. This image demonstrates the anterior displacement of the rectum, the vagina, the uterus and the urinary bladder. The cystic mass is seen in close relation with the anterior surface of the coccyx, but no evidence of invasion of the surrounding structures was detected.

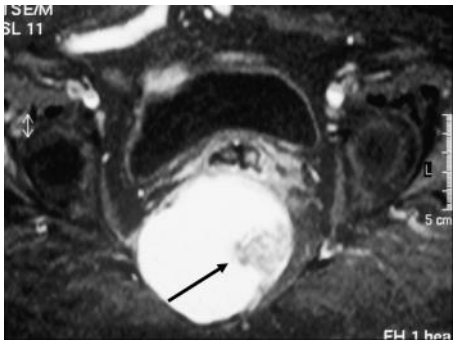

Fig. 2e - Axial fat-suppressed T1-weighted scan after gadolinium administration. The solid parietal vegetation strongly enhances (arrow)

Figure 3 Fig. 3 - Gross specimen.

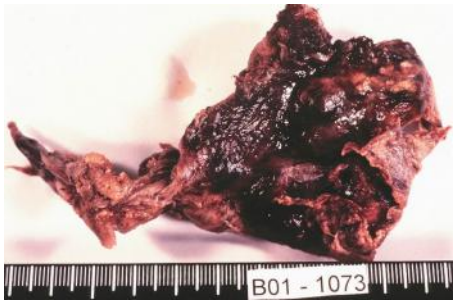

Fig. 3a - Gross specimen. A 11cm cystic tumor with coccyx - cystic tumor inner surface.

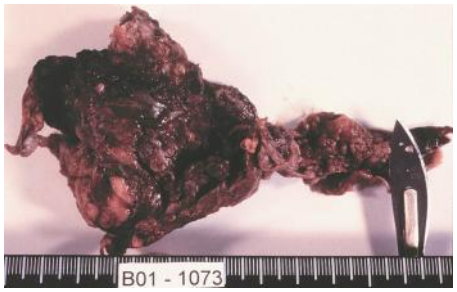

Fig. 3b - Gross specimen. A 11cm cystic tumor with coccyx - cystic tumor outer surface.

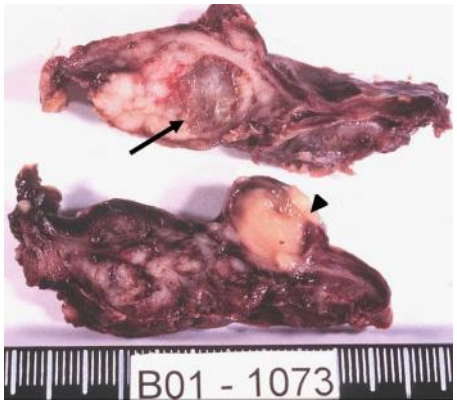

Fig. 3c - Gross specimen. Macroscopic aspect of wall section showing a solid hemorrhagic area (arrow) and a small cavity containing fat (arrowhead).

Figure 4 Fig. 4 - Histological section

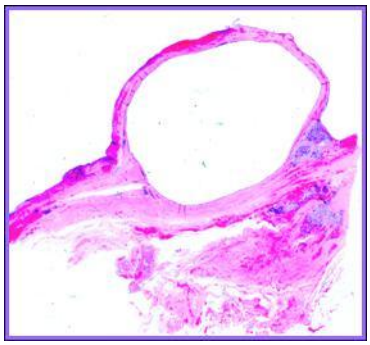

Fig. 4a - Histological section showing a squamous cell epithelium covering the small cystic cavity after removing the content (H&E).

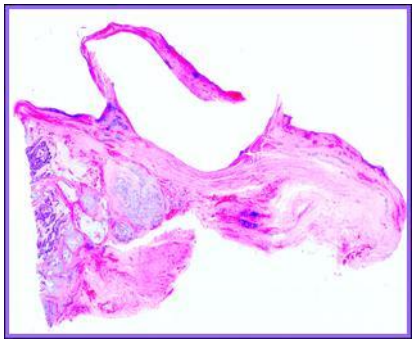

Fig. 4b - Histological section showing a squamous cell epithelium covering the small cystic cavity after removing the content (H&E).

Figure 5 Fig. 5 - Microscopic view.

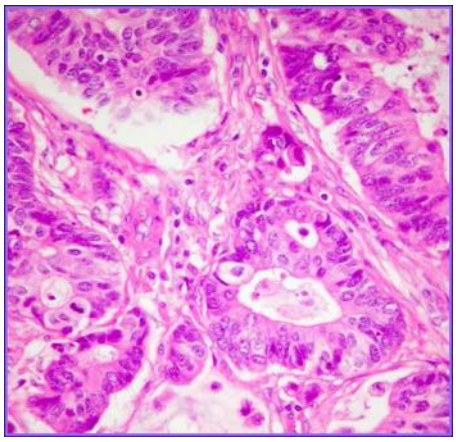

Fig. 5 - The solid area identified on MRI corresponds to an adenocarcinoma arising in a teratoma (H&E).

MeSH

Teratoma [C04.557.465.910]

A true neoplasm composed of a number of different types of tissue, none of which is native to the area in which it occurs. It is composed of tissues that are derived from three germinal layers, the endoderm, mesoderm, and ectoderm. They may be solid or cystic and are classified histologically as mature, immature, and malignant. (From Dorland, 27th ed & DeVita Jr et al., Cancer: Principles & Practice of Oncology, 3d ed, p1642)

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Sacrococcygeal Teratoma with malignant transformation {Online}
URL: http://www.eurorad.org/case.php?id=4366