Sacrococcygeal teratoma [SCT] in adults: Report Of A Case And Literature Review

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Citation

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Abstract

Background: Sacrococcygeal teratomas [SCT] are derived from embryonic germ-cell layers and are mostly encountered in infants. In adults, this entity is extremely rare and always worrisome for malignancy.
Methods: The authors present a case of a 42-year-old woman with the pathologic diagnosis of SCT. The clinical presentation, the histologic findings and the patients' clinical outcome are described. A review of the literature on SCT in adults is also presented.
Results: In this patient, who had no evidence of recurrence after adequate resection, examination of the specimen showed mature SCT.
Conclusion: Although rare in adults, SCT should be considered in the differential diagnosis of patients with a pelvic mass presenting with obstructive symptoms. Modern imaging techniques may be helpful to delineate the extent of the mass, and surgical removal is generally indicated at the time of detection. Sacrococcygeal mature teratoma is surgically curable if the lesion is completely resected. The presence of malignant transformation is associated with a less favorable outcome.

SUMMARY

The sacrococcygeal area is the most frequent site of teratoma in infancy. This tumor rarely presents in adulthood, and is confined mostly to the intrapelvic space. Most common are benign, also called mature teratomas. We report a case of adult saccroccocygeal teratoma treated in our hospital, and a literature overview.Introduction

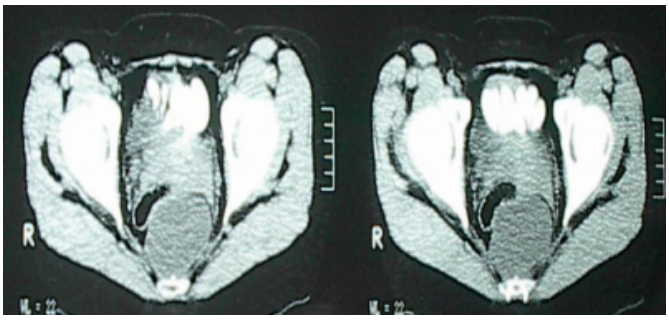
The sacrococcygeal area is the most frequent site of teratoma in infancy [1]. A female, rather than male, predominance exists [2,3]. This tumor rarely presents in adulthood. Unlike teratomas in infants, which are externally visible in 90% of cases, sacrococcygeal teratomas in adults are confined mostly to the intrapelvic space [4]. Most common are benign, also called mature teratomas. Rare cases of adults with malignant teratomas have been reported. We report a case of adult saccroccocygeal teratoma treated in our hospital. A literature overview of this entity with emphasis on adults is also presented.

CASE STUDY

A 42-year-old woman with a history of gradually increasing low back pain and constipation was referred for management of a soft tissue tumor of unknown origin, in September 2002. The mass had been identified in the presacral area on computed tomography SCAN [Figure 1] in another hospital.

Figure 1

Figure 1: CT scan showing a mass in the presacral area



The mass was originally detected and biopsied one year earlier, showing gelatinous, non malignant material. An attempt to remove the tumor at the time of the original diagnosis, through a laparotomy, failed due to excessive bleeding and fixation of the mass. Although fine-needle aspiration was negative for malignancy and the lesion remained stable on subsequent imaging studies, the patient continued to complain for low abdominal pain, constipation, and sensation of incomplete emptying of the rectum. Physical examination was normal except for a large, soft presacral mass felt on rectal examination. Neurological examination was with no abnormal findings. Alpha-

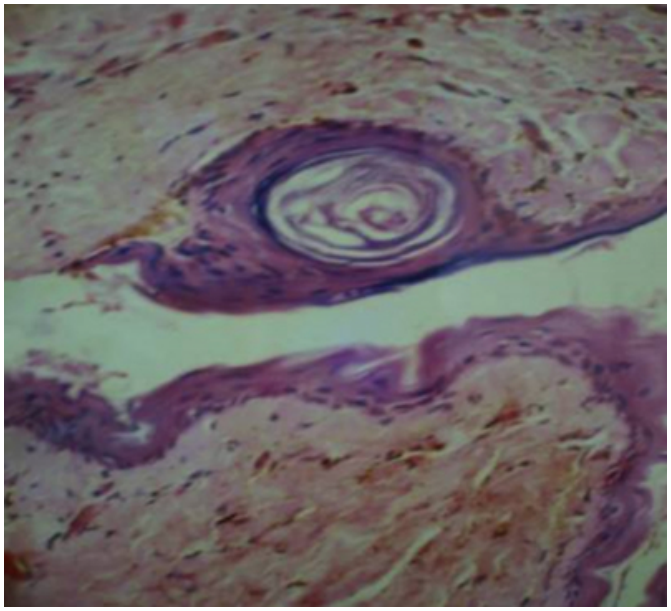
fetoprotein, carcino-embryonic antigen and human chorionic gonadotropin levels were normal. Considering the history and the clinical findings, the surgical removal of the smooth, tense-elastic presacral-extramucosal mass was recommended.

A laparotomy was performed, and the tumor was removed in toto, with an estimated blood loss of 350cc. There were no postoperative complications.

On gross examination, the specimen weighed 94g and measured 9.0x7.2x3.4cm. The cyst revealed a pink inner surface with slightly granular appearance and foci of yellow-brownish tissue of 1.0-1.8cm in maximum dimension. On microscopic examination, teratomatous elements, including ectodermal elements, were seen [Figure 2].

Figure 2

Figure 2: Microscopic examination of the excised mass

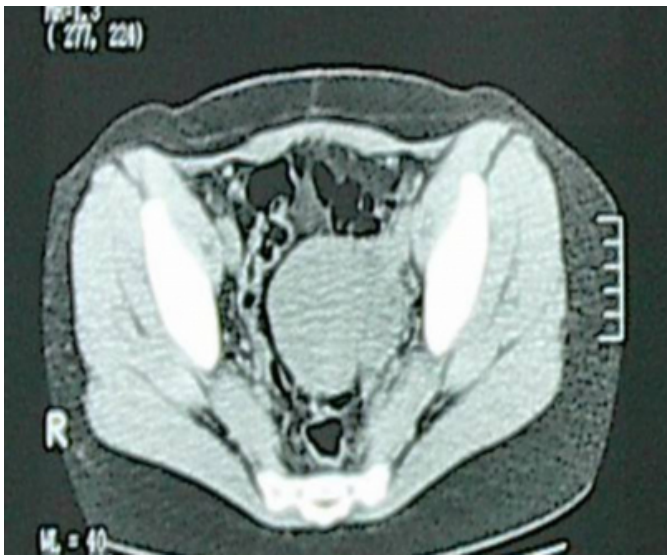


No malignant tissue of germ cell origin or malignant, nongerm cell elements, including adenocarcinoma and squamous cell carcinoma, were found.

Although the histologic findings showed a mature teratoma [also known as benign teratoma], and complete resection was accomplished, a full physical examination was performed periodically, with emphasis on assessment of the perineal and presacral area by rectal examination. One year later, CT scan of the pelvic area, showed no evidence of recurrence [Figure 3]. Within 3 years of follow-up, the patient had no evidence of recurrence.

Figure 3

Figure 3: CT scan of the same patient one year postoperatively



DISCUSSION

Sacrococcygeal teratomas and anal duct or anal gland cysts are the major cysts found in the perianal region [5]. The sacrococcygeal area is the most frequent site of teratoma in infancy, occurring in 1 of 35,000-40,000 live births [6]. A female, rather than male [4:1], predominance exists [2,3]. This tumor rarely presents in adulthood. Unlike teratomas in infants, which are externally visible in 90% of cases, sacrococcygeal teratomas in adults are confined mostly to the intrapelvic space [4]. Little is known about the embryogenesis of SCT, but it has been hypothesized that the anomaly is derived from a primitive knot, also referred to as Henson's node [7]. SCTs are thought to originate from multipotential cells in Henson's node, which migrates caudally to rest in the coccyx. They may expand postero-inferiorly into the gluteal area and/or antero-superiorly into the abdominopelvic cavity[8]. Various other theories exist to explain the origin of SCT. These include nonsexual reproduction of germ cells within the gonads or in extragonadal sites; ‘wandering' germ cells of non-parthenogenetic origin left behind during the migration of embryonic germ cells from yolk sac to gonad; or origin in other totipotential embryonic cells [9]. There is a tendency among the pediatric population toward malignant transformation of SCTs with increasing age [6]. However, in adult patients benign tumors predominate [10].

The first instance of SCT in an adult was described in 1847 by Emmerich [11]. Reviews of the literature by Head et al. [12], Ahmed and Pollock [1], Ng et al. [13], Bull et al. [14],

Miles and Stewart [2], and case reports published by other authors reveal a total number of 92 sacrococcygeal teratomas in adults, with only 20 cases described as malignant or with malignant transformation [15,16,17,18,19,20,21,22,23,24,25,26,27,28,29,30,31,32,33,34,35,36,37,38,39]. Patients with SCT may be asymptomatic on initial presentation, with the tumour discovered on rectal examination during routine physical examination or they may manifest a variety of symptoms that are not indicative of either benign or malignant tumours. Especially, if the mass is large enough, patients may complain of constipation due to rectal obstruction or recurrent urinary tract infections due to obstruction of the bladder neck [27,37]. If a SCT has directly invaded the nerve roots of the cauda equina or metastasized to the spinal cord, the patient may complain of neurologic symptoms such as lower-extremity numbness or weakness. However, this is very rare [9].

Sacrococcygeal teratomas and simple cysts account for the majority of cystic lesions in the presacral space but several other differential diagnoses have to be taken into consideration, such as anterior meningocele, rectal or anal duplication cyst, anal gland cyst, seroma, urinoma, tail gut cyst [retrorectal cystic hamartoma], chronic retrorectal abscess, pilonidal or dermoid cyst, soft tissue or bone tumours, osteomyelitis of the sacrum, chordoma, neurofibroma, fibrosarcoma, giant cell tumour or sacrum, postinjection granuloma and tuberculosis [2,40]. Serum tumor markers, such as alpha-fetoprotein and human chorionic gonadotropin, are not helpful in differentiating between benign and malignant lesions, but could be used in selected cases for postoperative detection of recurrences [13]. Invasion of adjacent structures, rather than simple displacement, sacral destruction and secondary findings such as locoregional lymph node and distal metastases are clearly indicative of malignancy [40,41].

SCTs, are classified into 3 histopathologic categories: (1) mature, (2) immature, and (3) malignant [42]. Mature teratomas (also known as benign teratomas) contain obvious epithelial-lined structures, mature cartilage, and striated or smooth muscle. Immature teratomas have areas of primitive mesoderm, endoderm, or ectoderm mixed with more mature elements in a highly cellular stroma with mitotic figures. Malignant teratomas, in addition to mature and/or embryonic tissues, have frankly malignant tissue of germ cell origin, such as germinoma and choriocarcinoma. Tumors containing malignant, non-germ cell elements, including adenocarcinoma and squamous cell carcinoma, are referred to as teratoma with malignant transformation. Patients with either a malignant teratoma or a benign teratoma with malignant transformation have a considerable increase in mortality, dying from the disease within 2 months to 2 years, whereas patients with benign disease, can live free of disease for more than 4 years after treatment [2].

Complete surgical excision is the treatment of choice. Depending on the size and topographic location of a lesion, either a transacral, transabdominal, or a combined approach could be performed [43,44]. In a case of sacrococcygeal teratoma, it is recommended that the coccyx should also be removed, because failure to remove it has been associated with a high risk of recurrence [2,9]. Solid teratomas may be very vascular, causing important intraoperative heamorrhage [2,43]. In the presence of larger tumors pre-operative angiography may be considered for blood supply evaluation and embolisation [7,40]. For histologically benign teratomas, adequate surgical excision is virtually curative. For malignant teratomas, surgical excision alone is inadequate, and patients should receive additional treatment with chemotherapy and/or radiotherapy. However, the role of adjuvant chemotherapy and radiotherapy is uncertain and has not proved to be beneficial in teratomas with malignant transformation; therefore, surgical extirpation remains the best therapeutic option for such cases [45]. Newer protocols, however, that include cisplatin, bleomycin, vinblastine, and radiotherapy may improve the results in the future [46]. For sacrococcygeal teratomas, postoperative outpatient follow-up is crucial. If complete resection is accomplished, a full physical examination should be performed periodically, with emphasis on assessment of the perineal and presacral area by rectal examination. CT scan or MRI may be useful if a recurrence is suspected.

In conclusion, although rare in adults, sacrococcygeal teratoma should be considered in the differential diagnosis of patients with a pelvic mass presenting with obstructive symptoms. Surgical removal is generally indicated at the time of detection, as these lesions carry a significant malignant potential.

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