Adult sacrococcygeal teratomas

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ABSTRACT

Sacrococcygeal (SCT) teratoma are rare in adults with an incidence of 1:87,000 and a female to male ratio of 10:1. Most of SCT are cystic and benign. Our patient was born with this mass that increased in size as she grew older. Being a precious child she was kept away from seeking medical advice by her parents. Initial work up included plain x-ray pelvis, computerized tomography scan and magnetic resonance imaging. At operation the mass was seen arising from sacrococcygeal region. It was excised completely along with coccyx. Pathologically, the specimen contained differentiated tissue from all 3 germ layers.


Case Report. A 22-year-old lady presented with huge mass at left gluteal region (Figure 1). This was present from birth and increased in size, as she grew older. Other than social embarrassment she only experienced pain in the gluteal area especially on sitting. She was married and had 2 children born by normal vaginal delivery. She was medically fit and had no neurological symptoms. Family history was unremarkable. Her physical examination showed mass at left gluteal region measuring approximately 20 x 20 cm extending to right gluteal region. Surface was smooth and irregular with prominent veins. It was non-tender, firm to hard in consistency, non-pulsatile and non-compressible. Neurovascular examination was unremarkable. Her base line laboratory work up included alpha fetoprotein which was normal.

Plain x-ray on the pelvis showed abnormal calcification on the left side of pelvis over the pubic bone. Computed tomography revealed non-enhancing pelvic mass with intrapelvic and external components. The intrapelvic part was on the left side of the pelvis inferiorly and the external part was posteriorly at gluteal region. It seemed to originate from the tip of the coccyx. It was a mixture of bone, cyst and small solid component with fat predominating. Bony component was noted...
Adult sacrococcygeal teratomas ... Al-Essa et al

Figure 1 - A huge mass about 20 x 20 cm occupying the left gluteal area extending to the right gluteal region. Surface is irregular with firm to hard in consistency. Prominent veins can be seen on the surface.

Figure 2 - Computed tomography revealing about 20 x 20 cm non-enhancing pelvic mass with intrapelvic and external components. The intrapelvic part is seen on the left side of the pelvis inferiorly and the external part is seen posteriorly at gluteal region. It seemed to originate from the tip of the coccyx. It is a mixture of bone, cyst and small solid component with fat predominating. Bony component is noted posteriorly.

Posterior sacral approach was used for the excision of tumor. At operation, the tumor was found to be multi lobulated, originating from the sacrococcygeal region. It was not involving the major neurovascular bundles. Tumor was excised completely along with coccyx. Soft tissues and neurovascular structures were well preserved. Postoperative period was uneventful.

Macroscopic examination showed a large soft tissue tumor weighing 2000 grams, measuring 25 cm in diameter predominantly fat with an irregular portion of bone and cystic areas. The largest cyst was 4 cm in diameter containing yellow greasy material and hair. Microscopically, it was a mature benign cystic teratoma exhibiting lobules of mature adipose tissue with areas of muscles, fibrous tissue and blood vessels. Irregular cystic spaces lined by stratified squamous epithelium with adjacent sebaceous glands were seen. Other cysts were lined by low cuboidal epithelium, gastric and nasal mucosa. Extensive areas of fat necrosis were noted. No immature elements or malignancy were seen.

**Discussion.** Sacrococcygeal teratoma are rare congenital tumors that develop embryologically from multipotent cells in Hensen’s node and enlarge as pre or post sacral mass. All SCTs involve the coccyx. It is usually diagnosed in infancy. It occurs on 1.40,000 births with female predominance of 4:1.1 There is a risk of malignancy which increases with age. Neonates have a 2-5% risk of malignancy that increases to 50% at one year and nearly 100% after 5 years of age.2 The incidence of malignancy in pediatric SCT increases directly with age, intrapelvic component and the ratio of solid to cystic tissue, but those that persist into adulthood are usually benign. Cystic and well differentiated tumors with mature elements tend to be benign, solid tumors with embryonic elements are usually malignant. Exceptionally these tumors are observed in adults with incidence of 1:87,000 and female to male ratio of 10:1.3 Most adult SCTs are cystic, and only 1-2% are malignant. The familial SCT has higher incidence in twins and associated with congenital abnormalities, especially of the spine.4 A triad of anal stenosis, sacral dysplasia and presacral mass has been identified in several families as well. The familial SCT tends to be benign and well adherent to the rectum.5 Mature SCT can undergo dysplastic change and frank malignant degeneration.6 Malignancy in an SCT usually arises from a single germ line and generally develops from embryonic tissues. Serum levels of the fetal oncogenes alpha fetoprotein, carcinoembryonic antigen and human chorionic gonadotrophin are elevated in patients with malignant SCT.7 The symptoms of SCT may include thin stools, constipation, urinary frequency, low back pain, lower extremity paresthesia or paraparesis and bilateral venous engorgement of the lower extremities.8 Pelvic and rectal examination may elicit a mass, extrinsic compression of the vagina, displacement of the uterus, displacement or extrinsic compression of the posterolateral rectum, erosion or protrusion into the rectum or perirectal posterior mass, dimple or fistulous tract.9 Differential diagnosis of SCT in adults includes chordoma, ependymoma, giant cell tumor of sacrum, perirectal
In conclusion, SCTs are rare in adults and most of them are benign and cystic and almost always involve coccyx. Plain x-ray on the pelvis, CT or MRI and alpha fetoprotein are the diagnostic modalities that can also be used for follow up. Recurrence of benign SCT is very rare if coccyx is excised completely along with the tumor. Malignant tumor have high recurrence rate.

References