Adult sacrococcygeal teratomas

Abdul-Aziz Al-Essa, FRCS (Glas), FRCS (Edin), *Tauqeer A. Malik,* MBBS, MRCS Ed, *Mohammad K. Baghdadi,* MBBS, DIS, *Ali A. El Tayeb,* DCP, FCS(Path).

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.

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Adult sacrococcygeal teratoma (SCT) is extremely rare in adult with an incidence of 1:87,000 and a female to male ratio of 10:1. Sacrococcygeal teratoma are congenital tumor that develop embryologically from multipotent cells and enlarge as pre and post sacral mass. All SCTs involve the coccyx. Most of the SCTs are cystic and benign and only 1-2% are malignant. The risk of malignancy increases with age. Familial SCT has been identified and is associated with congenital abnormalities especially of spine. The symptoms are usually due to the pressure on surrounding structures. The investigations of choice are plain x-ray, computerized tomography (CT) and magnetic resonance imaging (MRI). Biochemical markers including alpha fetoprotein, carcinoembryonic antigen and human chronic gonadotrophin are also helpful. Surgical treatment of SCT is complete excision, inclusive of coccyx and may be sacrum if involved. Various surgical approaches have been defined. The recurrence rate of SCT is 7.5-22% which can reach up to 37% if coccyx is not excised.

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

From the Department Surgery, King Fahd Armed Forces Hospital, Jeddah, *Kingdom of Saudi Arabia*.

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Address correspondence and reprint request to: Brig. Gen. (Dr.) Abdul Aziz Al-Essa, King Fahd Armed Forces Hospital, PO Box 9862, Jeddah 21159, *Kingdom of Saudi Arabia*. Tel. +966 (2) 6653000 Ext. 3038. Fax. +966 (2) 6652469. E-mail: surgery@kfafh.org

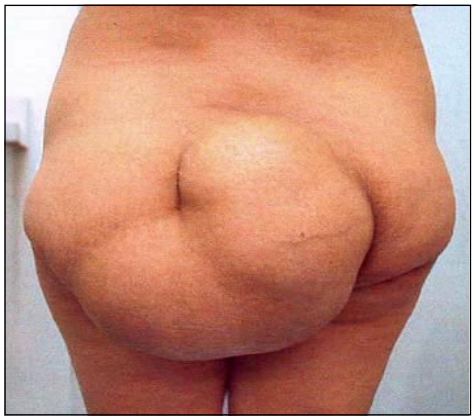

Figure 1 - A huge mass about 20 x 20 cm occupying the left gluteal area extending to the right gluteal region. Surface is smooth irregular with firm to hard in consistency. Prominent veins can be seen on the surface.

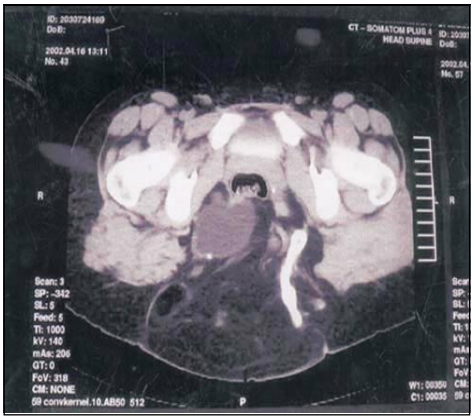

Figure 2 - Computed tomography revealing about 20 x 20 cm non-enhancing pelvic mass with intrapelvic and external components. The intrapelvic part is seen on the left side of the pelvis inferiorly and the external part is seen posteriorly at gluteal region. It seemed to originate from the tip of the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

posteriorly (**Figure 2**). 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.[6] Differential diagnosis of SCT in adults includes chordoma, ependymoma, giant cell tumor of sacrum, perirectal

abscess, rectal tumor, fibrosarcoma, neurofibroma or pilonidal cyst. If SCT is suspected, transrectal needle biopsy is contraindicated due to possible dissemination or leakage of a teratoma and subsequent recurrence, whether the tumor contains malignant components or not.[9] Surgical treatment of SCT is complete excision, inclusive of coccyx and may be sacrum if involved. Anterior abdominal, posterior sacral, perineal or combined approaches have been used successfully. If the tumor is less than 8-10 cms, whether complex or simple, the posterior sacral approach is appropriate. For larger tumors, the combined abdominosacral approach in the lateral position or abdominoperineal approach is used but latter is better in avoiding sacral nerve injury.[9] The initial control of the vascular supply of the tumor can avoid excessive intraoperative blood loss.[9] Preoperative angiography can be helpful to outline the tumor blood supply. Other complications may include sacral nerve, ureteral, rectal, and dural injury.[9] The recurrence rate of SCT has been reported to be 7.5-22% which can reach up to 37% if coccyx is not excised.[9] Most recurrences are within 2 years of the initial excision may be due to retained SCT tissue or spillage of cyst contents during dissection or due to residual tissue densely adherent to vital structures left behind at initial operation. Malignant tumors have a greater likelihood of recurrence and the incidence of malignancy is higher in recurrent than in primary tumors.

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.

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