

Sacrococcygeal –teratoma in adults: A case report and review of literature

Seheli Bandyopadhyay¹, Abhishek Basu², Arnab Bhattacharjee³, Arijit Sen⁴,
Soumadip Panda⁵, Swarnendu Datta⁶

¹Department of Radiotherapy, R.G.Kar Medical College and Hospital, Kolkata, India, ²Department of Radiotherapy, R.G. Kar Medical College and Hospital, Kolkata, India, ³Department of Radiotherapy, R.G. Kar Medical College and Hospital, Kolkata, India, ⁴Department of Radiotherapy, R.G. Kar Medical College and Hospital, Kolkata, India, ⁵Department of Radiotherapy, R.G. Kar Medical College and Hospital, Kolkata, India, ⁶Department of Radiotherapy, R.G. Kar Medical College and Hospital, Kolkata, India,

Abstract: Sacrococcygealteratoma (SCT) is known as a disease of infants but is rare in adults.. This is an uncommon location of teratoma in adults and malignant transformation has been rarely described. The clinical, radiological and histopathological characteristics of this case are described. Although complete surgical resection is the standard of care, with malignant transformation SCTs have a poor outcome. We report here the case of a 53 yr old female with sacrococcygeal teratoma

Key words: Adult, Sacrococcygeal, teratoma

I. Introduction:

Sacrococcygeal teratoma (SCT) is common in infancy and childhood, with an estimated prevalence of 1 in 35,000–40,000 live births [1]. They can be diagnosed prenatally by fetal ultrasound and 50–70% are found during the first few days of life. [1,2]. It occurs more often in girls; with female to male ratio of 3-4:1.[3-6] Most of the SCTs are cystic and benign and only 1-2% are malignant.[7] The cysts may be filled with serous fluid, mucoid, or sebaceous material and lined by true epithelium. Radiological imaging is helpful in the diagnosis of these lesions and in delineating their extent. Reported cases of sacrococcygeal teratomas in adults are rare. Reviews of the literature by Head et al [8], Ahmed and Pollock [9], Ng et al [10] and Bull et al [11] Luk et al [12], Miles et al [13] found a total of 92 reported cases, with only 16 described as malignant or with malignant transformation. Most SCTs are benign, also called mature teratomas. Rare cases of adults with malignant teratomas have also been reported. Unlike teratomas in infants, which are externally visible in 90% of cases, sacrococcygeal teratomas in adults are confined mostly to the intrapelvic space [14]. Treatment consists of complete surgical resection but incomplete surgical removal carries a poor prognosis.

We report a case of adult sacrococcygeal teratoma treated in our hospital. The patient was a 53-year-old woman presenting with intestinal and urinary obstructive symptoms due to a large sacrococcygeal teratoma with malignant transformation. Imaging appropriately demonstrated the combined cystic and solid nature of the tumour, its extension and relations with adjacent structures. A literature overview of this entity is also presented.

II. Case Report

A 53-year-old lady presented with gradual onset constipation and low back pain for one year which gradually increased in intensity. She had had two episodes of urinary retention recently. On gynaecological and rectal examination a large palpable firm mass was found in the right hemipelvis, posterior to the rectum. Neurological examination was unremarkable. Alpha-fetoprotein (AFP), Carcinoembryonic antigen (CEA), Human chorionic gonadotropin (β – hCG), CA 125 and Lactate dehydrogenase (LDH) levels were normal.

CT scan showed a complex heterogeneously enhancing lesion with cystic changes in the pelvis extending posterior to the sacrum and eroding it. The lesion was ill-defined in outline with streaky opacities around it and measured 11 × 10 × 8 cm. Rectum and sigmoid colon were displaced towards left side. Although the mass displaced the uterus, adnexae and urinary bladder anteriorly, it was clearly separated from them. Few calcifications were seen in the capsule of the cyst.

The patient underwent bloc surgical removal of the pelvic mass along with total abdominal hysterectomy and bilateral salphingoophorectomy. Histopathology revealed well-differentiated adenocarcinoma arising in a sacrococcygeal teratoma (Fig 1a, 1b, 2).

She was referred to our institute after eight months with symptoms of pelvic pain and urinary retention. Contrast-enhanced CT Scan revealed a large, irregular, ill-defined, infiltrating mass lesion measuring 5 × 5.5 × 6

cm in presacral space extending to retrosacral region eroding sacrum and coccyx (Fig 3). The lesion had septated cystic component with irregular enhancing solid tissue. Anteriorly the lesion had extended to bladder and urethra and posteriorly to medial wall of rectum with loss of intervening fat plane. The lesion has also infiltrated bilateral pyriformis and right obturator internus muscles.

As the patient refused further surgical extirpation, she received External Beam Radiotherapy with ⁶⁰Cobalt to a dose of 60Gy in 30 fractions over 6 weeks with four field box technique and CT based planning. This period of treatment was uneventful with occasional mild episodes of diarrhoea which was controlled conservatively. She then received adjuvant combination chemotherapy with Inj Bleomycin, Inj Etoposide and Inj Cisplatin for three cycles (BEP regime). No significant chemotherapy induced toxicity was encountered except neutropenia which was managed by filgrastim support. After three months of completion of treatment (post radiation and chemotherapy) she had multiloculated cystic lesion with solid components in the presacral region extending to perirectal space indicating persistence of the disease (Fig 4). Further surgical salvage or second line chemotherapy was discussed with the patient but she refused any further active treatment. She was eventually offered best supportive care including palliation of pain and died after 2 months subsequently.

III. Discussion

Teratomas are germ cell tumours commonly composed of multiple cell types derived from one or more of the three germ cell layers. They contain tissue foreign to their anatomic site and not resulting from metaplasia. They may be inherently malignant or have the potential for malignant degeneration. Many theories of origin have been postulated for teratomas including parthenogenetic development of germ cells within the gonads or in extragonadal sites; “wandering” germ cells of nonparthenogenetic origin left behind during the migration of embryonic germ cells from yolk sac to gonad; or origin in other totipotential embryonic cells. Sacrococcygeal teratomas (SCT) are tumours that arise primarily from this region and, the sacrococcygeal area is the most frequent site in infants and it is common in the intrapelvic region in adults. SCTs are rare in adults with only a few published cases in the literature.[8-13]

Though 90% of SCT in neonates are externally visible, most of the adult SCT present as intrapelvic masses. They may be asymptomatic in the initial phase but symptoms appear as the mass increases in size and are related to the mass effects or bulk of the tumour. Patients may complain of low back pain, chronic fistula, or obstructive symptoms of the gastrointestinal or genitourinary tracts with bowel or urinary symptoms like constipation or recurrent urinary tract infection or urinary retention. Venous engorgement of the lower limbs and lower extremity motor power losses may also be seen.

Roentgenogram revealing calcifications in the coccygeal region or an anterior displacement of the rectum in the barium enema suggests SCT[13]. Computed tomography and MRI are the most significant tools to define the mass of SCT, to evaluate the intrapelvic extension and relationship to other structures. Most commonly, teratomas appear as a complex mass with roughly equal amounts of solid heterogeneous and cystic areas with or without septations. They also frequently present as thick walled cystic masses, sometimes multiloculated, that may contain fat, calcified elements and/or small solid nodules. Complex, predominantly solid tumours with significant areas of necrosis within the tumour are more likely to be malignant. Invasion of adjacent structures, sacral destruction and secondary findings such as locoregional lymph node and distant metastases are clearly indicative of malignancy.[1,2]

Microscopically, teratomas usually contain a variety of tissues from more than one germ layer. The teratomas are classified into three histopathologic categories: mature, immature, and malignant. Mature teratoma (also referred to as benign teratoma) contains obvious epithelial-lined structures as well as mature cartilage and striated or smooth muscle. Immature teratoma has areas of primitive mesoderm, endoderm, or ectoderm mixed with the more mature elements. The presence of a highly cellular stroma exhibiting mitotic figures is sufficient to make this diagnosis. Malignant teratomas have frankly malignant tissue of germ cell origin, such as embryonal tissue, germinoma (seminoma or dysgerminoma), yolk sac tissue, and choriocarcinoma, in addition to mature and/or embryonic tissues.[15]

Differential diagnoses of these masses include anterior meningocele, rectal duplication cysts or anal gland cysts, chordoma, meningocele, neurofibroma, fibrosarcoma, giant cell tumour of sacrum, pilonidal cysts, osteomyelitis of sacrum, fistula with presacral extension and abscess formation, postinjection granuloma, and tuberculosis.[13] The standard treatment for all primary SCTs is surgical excision. As these tumours are contiguous with the coccyx, it is recommended that the coccyx be removed. Failure to remove the coccyx may be associated with a high risk of recurrence[3]. If the tumour is histologically benign or immature teratoma without frank malignant tissue, complete excision is adequate. For malignant teratomas, surgical excision alone is inadequate and patients should receive additional treatment with chemotherapy and/or radiotherapy. As adult SCT is rare, there has been no standard recommendation for the use of chemotherapy or radiation.

Fig 3: Contrast enhanced CT Scan revealed a large, irregular, ill-defined, infiltrating mass measuring $5 \times 5.5 \times 6$ cm in presacral space extending to retrosacral region.



Fig 4: Post radiation and chemotherapy CECT Scan after three months of completion of treatment revealed multiloculated cystic lesion with solid components in the presacral region extending to perirectal space indicating persistence of the disease

IV. Conclusion:

Although a rare tumour per se and even rarer in adults, sacrococcygeal teratoma must be considered in the differential diagnosis of all intrapelvic solid – cystic masses in adults, especially if they are presacral in location. Complete surgical excision and benign pathology have a good prognosis but malignant pathology and incomplete excision have poor survival and outcome. Our report underlines the importance of being vigilant and suspicious in such rare cases to enable timely diagnosis and proper intervention.

References

- [1]. Keslar PJ, Buck JL, Suarez ES. *Germ cell tumors of the sacrococcygeal region: radiologic-pathologic correlation. Radiographics 1994;14:607–20.*
- [2]. Wells RG, Sty JR. *Imaging of sacrococcygeal germ cell tumors. Radiographics 1990;10:701–13.*
- [3]. Schropp KP, Lobe TE, Rao B, Mutabagani K, Kay GA, Gilchrist BF, et al. *Sacrococcygeal teratoma: The experience of four decades. J Pediatr Surg 1992;27:1075–8.*
- [4]. Pauniah SL, Heikinheimo O, Vettenranta K, Salonen J, Stefanovic V, Ritvanen A, et al. *High prevalence of sacrococcygeal teratoma in Finland -A nationwide population-based study. Acta Paediatr 2013;102:e251–6.*
- [5]. Golas MM, Gunawan B, Raab BW, Füzesi L, Lange B. *Malignant transformation of an untreated congenital sacrococcygeal teratoma: A amplification at 8q and 12p detected by comparative genomic hybridization. Cancer Genet Cytogenet 2010;197:95–8.*
- [6]. Swamy R, Embleton N, Hale J. *Sacrococcygeal teratoma over two decades: Birth prevalence, prenatal diagnosis and clinical outcomes. Prenat Diagn 2008;28:1048–51.*
- [7]. Al-Essa AA, Malik TA, Baghdadi MK, El Tayeb AA. *Adult sacrococcygeal teratomas. Saudi Med J 2004;25:3679.*
- [8]. Head H, Gerstein J, Muir R. *Presacral teratoma in the adult. Am Surg 1975;41:240–8.*
- [9]. Ahmed HA, Pollock DJ. *Malignant sacrococcygeal teratoma in the adult. Histopathology 1985;9:359–63.*
- [10]. Ng EW, Porcu P, Loehrer PJ. *Sacrococcygeal teratoma in adults: case reports and a review of the literature. Cancer 1999;86:1198–202.*
- [11]. Bull J Jr, Yeh KA, McDonnell D, Caudell P, Davis J. *Mature presacral teratoma in an adult male: a case report. Am Surg 1999;65:586–91.*
- [12]. Luk SY, Tsang YP, Chan TS, Lee TF, Leung KC. *Sacrococcygeal teratoma in adults: Case report and literature review. Hong Kong Med J 2011;17:417–20.*
- [13]. Miles RM, Stewart GS Jr. *Sacrococcygeal teratomas in adult. Ann Surg 1974;179:676–83.*
- [14]. Ravitch MM, Smith EI. *Sacrococcygeal teratoma in infants and children. Surgery 1951;30:733–62.*
- [15]. Mahour GH. *Sacrococcygeal teratomas. CA Cancer J Clin 1988;38:362–7.*