Original Article

Case report
Sacrococcygeal teratoma with yolk sac elements (endodermal sinus tumors) in two years old child

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Summary:

Background: Sacrococcygeal teratoma is seen in 1 in every 35000 live births, and is the most common tumor presenting in newborn and children but also reported in adults.(1)
Sacrococcygeal teratoma are the most common type of germ cell tumors (both benign and malignant) diagnosed in neonates , infants and children younger than 4 years.(2)
Case report: A 2 years old male child presented with a visible lump or mass under the skin at the top of the buttocks crease after falling on the ground on his buttocks, and the parents give a history that the child had a constipation since birth.
The case was diagnosed as endodermal sinus tumor with yolk sac elements on the bases of clinical examination, ultrasonography, CTscan and fine-needle aspiration cytology.
After diagnoses patient referred to the oncologist for multi-drug chemotherapy and surgical removal by the pediatric surgeon after shrinkage of the tumor mass.
Conclusion: Infants and young children present with a palpable mass in the sacropelvic region have a greater likelihood of being malignant.
Keywords: endodermal sinus tumors, sacrococcygeal teratoma , yolk sac tumor

Introduction:

Sacrococcygeal germ cell tumors in neonates and infants are nearly always primary. (3) From 75%-90% of the cases occur in females. They exhibit 2 peaks in incidence : the first approximately 2 years of age represents congenital neoplasms , the second in late adolescence.(4) In some cases of sacrococcygeal teratoma, there is an association with twinning or malformations.the most common location in descending order of frequency are the sacrococcygeal area , head and neck, retroperitoneum , mediastinum and central nervous system.(10) The most common type is the mature teratoma (approximately 75% of these tumor)presenting at birth in the sacrococcygeal region or protruding through the abdominal cavity .it may be very large , is usually cystic and multilocular and may appear malignant to the surgeon because of its adherence to the neighboring structures . This fixation is of an inflammatory nature. (5) About 12% are malignant and lethal, the remainder is immature teratoma and their malignant potential correlates with amount of the immature tissue, usually immature neuroepithelial elements present. Most of the clearly malignant teratomas have the appearance of Yolk sac (endodermal sinus) tumor, either pure or associated with other germ cell component, like embryonal carcinoma. (6, 7, 8). An interesting clinical observation is that the large majority of sacrococcygeal teratomas present at birth is benign, whereas tumor in the same location discovered after The age of 2 months is often malignant. (3, 4, 9) Regarding treatment and prognosis: most teratomas involve some combination of surgery and chemotherapy, before modern chemotherapy, this type of neoplasm was highly lethal, but the prognosis has significantly improved since 90% survival rate when treated effectively. (11, 12)

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Case report:

A 2 years old male child presented with a visible lump or mass under the skin at the top of the buttocks crease after falling on the ground on his buttocks, and the parents give a history that the child had a constipation since birth, referred to us in the teaching laboratory, the cytological department at Baghdad teaching hospital, for fine needle aspiration cytology. The ultrasonographic report reveals a pelvic mass in relation to the sacroccyx... CTscan exam showing a large retrorectal mass, infront of the sacrococcygeal area with contrast enhancement which is diagnostic of malignant teratoma. Fine needle aspiration under local anesthesia done with disposable plastic syringe, 10 ml (22-23gauge) . The aspirate smeared between 2 standard microscope slides, fixed in 95% ethyl alcohol, stained with Papanicolaou stain and examined by light microscope. The smears show large cells with vesicular, pleomorphic nuclei, pale nuclear chromatin but coarsely granular and irregularly distributed. Nucleoli are large eosinophilic, the cytoplasm stains variably and usually vacuolated, cell borders are indistinct. (Figure 1) Cells usually

form aggregates with a microglandular pattern and papillary clusters... (Figure 2) Fragments of highly cellular tissue composed of smaller spindle cells are found representing primitive mesenchymal tissue. (figure3). This cytological picture is highly diagnostic of endodermal sinus tumor (yolk sac tumor), which is malignant.

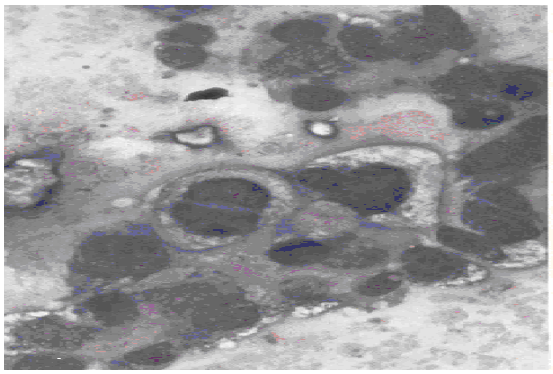

Figure 1: endodermal sinus tumors; showing loose cluster of large malignant epithelial cells with prominent cytoplasmic vaculations. (Pap stain,HP)

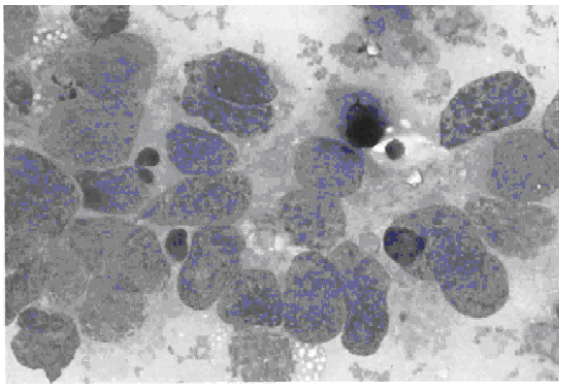

Figure 2: adenocarcinoma- like clusters of large malignant cells with large nuclei and very coarse chromatin. (Pap stain,HP)

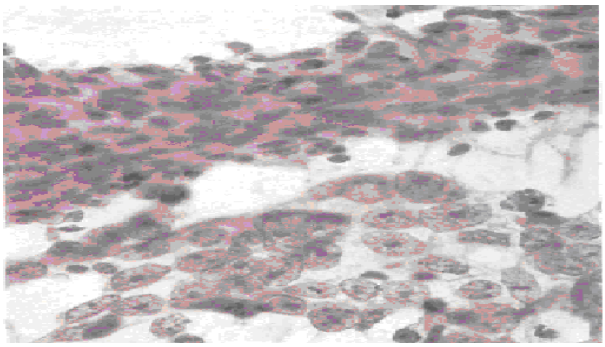

Figure 3: fragment of undifferentiated mesenchymal tissue (above); cluster of malignant epithelial cells;large vesicular nuclei;prominent large nucleoli (below).(Pap stain,H

Discussion and conclusion:

An interesting clinical observation is that the large majority of sacrococcygeal teratomas present at birth are benign , whereas tumors in the same general location discovered after the age of 2 months are often malignant.(6) This has been taken by some to indicate that a malignant transformation has supervened in that short period we doubt that this is the case. It seems to us that this clinical observation can better be explained by postulating the existence of 2 groups of teratomas. One arises in the very distant portion of the sacrococcygeal region, is therefore clinically obvious at the time of birth, and nearly always mature. The other arises more proximaly in the retrorectal or adjacent retroperitoneal region , is malignant from the start and grows in the sacrococcygeal area to become clinically evident only sometime after birth , this being responsible for the clinical observation that teratomas associated with marked bowel or bladder dysfunction are often malignant. (11, 12).

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