PEDIATRIC PATHOLOGY

Cavernous Hemangioma Presenting as a Right Adnexal Mass in a Child

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This is the case of an 11-year-old girl who presented with a right adnexal mass and vague abdominal symptoms since seven months prior to her hospital admission for surgery. CT-scan and sonographic images were those of a benign lesion, probably ovarian torsion or infarction. Serum tumoral markers were normal. A right salpingo-

oophorectomy and appendectomy were performed. Pathology examination revealed a cavernous hemangioma of the ovary. The clinicopathologic presentation of this unusual benign ovarian tumor is discussed.

Key words: Adnexa, Cavernous hemangioma, Ovary

emangiomas are fairly common pediatric lesions that may occur in any site or organ. Ovarian hemangiomas, however, are extremely rare, with no more than fifty documented cases reported in the medical literature (1-2). To our knowledge, only eight have been reported in the pediatric age (2). Most ovarian hemangiomas are discovered as an incidental finding or discovered during autopsy. Other cases have presented as ovarian masses (3-5), acute abdomen due to ovarian torsion (2,6-8), or ascites (4,6,7). The lesions are usually unilateral but bilateral ovarian hemangiomas have also been described (9). They have also been associated with hemangiomas at other sites, or with generalized hemangiomatosis and thrombocytopenia (9,10). We present the case of this girl who presented with vague abdominal symptoms and a right adnexal mass.

Case report

An eleven-year-old prepubertal girl was referred to our institution for a work up of malignancy. Her past medical history revealed a history of benign cardiac arrhythmias which required no medications, and gastro-esophageal reflux managed with Ranitidine as needed. She also related

an episode of a strong abdominal pain associated with bladder sphincter loss and dizziness seven months prior to admission. An abdomino-pelvic ultrasound study revealed a small, benign -looking ovarian cyst and no further follow-up was given at the time. Family history was negative for gynecological malignancies or bleeding diathesis. Two weeks prior to admission the patient had abdominal fullness sensation followed by a strong colicky pain, vomiting, and fever. The presence of hematuria on urinalysis, and increased white blood cell count prompted hospitalization. She received intravenous hydration, antifever measures and analgesics. She was discharged home but two days later returned to the hospital due to persistent abdominal pain and fever. An abdomino-pelvic CT-scan revealed a pelvic mass and the patient was referred to the University Pediatric Hospital for a complete work-up to rule out a malignancy.

At physical exam, the child was alert, cooperative and in no apparent distress. She had a soft, depressible abdomen with positive bowel sounds, and mild tenderness in the right lower quadrant. No peritoneal irritation was elicited and abdominal masses were not palpable. Small posterior cervical and left inguinal nodes were noted. Sexual development revealed a Tanner II. Her skin had no lesions. A pelvic sonogram showed a heterogeneous pelvic mass arising either from the cul-de-sac area or right ovary with evidence of fluid. An abdomino-pelvic CT-scan revealed a soft tissue mass measuring 7.0 x 5.5 cm over the right pelvic area displacing the uterus anteriorly and to the right.

Serum levels for alpha fetoprotein, human chorionic gonadotropin and carcino-embryonic antigen were within normal limits. A hemogram showed a white blood cell count of 9,900/mm³, hemoglobin of 11.4 gm/dl, platelet count of 519,000/mm³, and a normal differential count.

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Although radiographic images and protracted clinical course suggested a benign ongoing process, due to persistent abdominal pain, constitutional symptoms, presence of a large ovarian mass, and to establish a pathological diagnosis, the child was taken for surgery. Through a low transverse pelvic incision a right ovarian reddish swollen tumorous mass involving the fallopian tube and appendix was found. Free straw color fluid in the cul-de-sac was obtained. The peritoneal surface, retroperitoneal structures, liver, and the contralateral ovary had a normal appearance. After right salpingo-oophorectomy and appendectomy the child had an uneventful postoperative course.

The ovarian mass with attached segment of fallopian tube and appendix were received fixed in formalin. The specimen weighed 120 gm. The ovary was markedly enlarged and the fallopian tube distended, both presenting an external purplish discoloration. The ovary measured 8.5 x 5.5 x 4.0 cm, and the fallopian tube 6.0 cm in length and 1.4 cm in width. On sectioning the ovary, the cut surface was hemorrhagic with an area that was softer, spongy and irregular, that measured 4.2 x 3.5 cm (Figure 1). Only a 0.2 cm rim of gray tan soft tissue was seen at the periphery of this ovarian mass. Sections of the fallopian tube revealed a hemorrhagic content. The appendix was grossly normal.



Figure 1. Ovarian hemangioma. Cut surface is hemorrhagic with a spongy irregular appearance. Note distended fallopian tube (*) with hematosalpinx.

Microscopic sections of the ovary revealed a proliferation of large vascular channels lined by a single layer of endothelial cells. These vascular channels contained blood (Figure 2). Trichrome stain highlighted the vascular nature of this lesion. Immunostain for CD 31

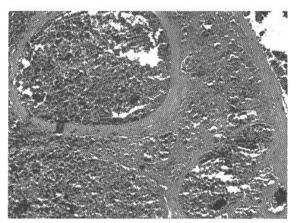


Figure 2. Note multiple dilated vascular channels with a blood content. 100X

revealed positive expression of endothelial cells supporting the vascular nature of the lesion. In addition, there was abundant hemorrhage, necrosis and dystrophic calcifications. Residual normal ovarian tissue was not identified. Sections of the fallopian tube showed a hematosalpinx and dystrophic calcifications. The appendix was unremarkable.

The pathologic diagnosis was that of an ovarian hemangioma, cavernous type, with extensive hemorrhages, necrosis, and dystrophic calcifications, and hematosalpinx most likely from torsion. Peritoneal washing for cytology examination showed reactive mesothelial cells with no evidence of malignancy.

Discussion

Ovarian lesions in children are most often benign cysts, follicular type, detected during sonography. Endometriosis, germ cell tumors, torsion of the ovary or massive edema of the ovary are additional rare lesions that may present as adnexal masses. Most benign tumors occur in prepubertal patients whereas malignant tumors peak after thirteen years of age. Torsion of the ovary and fallopian tube is considered a rare event in the pediatric population, and although some have reported that torsion almost always occurs secondary to a preexisting ovarian lesion, this is not the experience of most authors (11). In our case, the presence of a cavernous hemangioma in the ovary constituted the underlying pathology for the adnexal mass, causing torsion with hemorrhagic necrosis, calcifications, and hematosalpinx. This pathologic finding is in accord with the indolent clinical course of this patient, the absence of tumoral markers, and the radiographic images of a benign lesion.

Hemangiomas in the pediatric age are not so uncommon

and may occur in any site or any organ. Ovarian hemangiomas nonetheless are extremely rare, with no more than fifty documented cases reported in the medical literature (1-2). Of these, only eight have been reported in the pediatric age (2). The age of patients reported with ovarian hemangiomas ranges between 4 months and 63 years not showing predominance in any decade (1). Most cases are discovered as incidental findings or during autopsy. Other cases have presented as an ovarian mass (3-5), acute abdomen with ovarian torsion (2,6-8), or ascites (4,6,7). A recently reported case presented with an adnexal mass, ascites and elevated CA 125 (4). The lesions are usually unilateral but bilateral ovarian hemangiomas have been described (9). They have also been associated with hemangiomas at other sites, or with generalized hemangiomatosis, and thrombocytopenia (9, 10). Histologic types include cavernous, which is the most common, mixed capillary-cavernous type and cellular capillary type. The treatment of choice is surgery which in most cases result in complete cure.

Resumen

Presentamos el caso de una niña de 11 años con una masa en la aneja derecha y síntomas abdominales poco específicos desde siete meses antes de su hospitalización para cirugía. Las imágenes sonográficas y de CT demostraron una lesión benigna, probablemente una torsión o infarto de ovario. El nivel sérico de los marcadores tumorales fueron normales. Se realizó una

salpingooforectomía y apendectomía que demostraron al examen patológico un hemangioma cavernoso de ovario. Se discute la clínica y patología de este tumor benigno.

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313