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Anastomosing Hemangioma at the Root of Suspensory Ligament of the Ovary - A Case Report

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Abstract

Anastomosing hemangioma is an uncommon tumor of vascular origin usually restricted to the genitourinary tract. Here, we report an unusual presentation of this variant of hemangioma at the root of suspensory ligament of the ovary. Literature has evidence of few but sure cases of presentation of this tumor on the ovary also some cases which describe this lesion's predilection to soft tissues in the paraspinal area. The patient we received is a 53 year old female who had been diagnosed with a left pelvic mass about a month before coming to our department. Due to the location of the tumor on CT and MRI possibility of a schwannoma or an adnexal sex cord stromal tumor was considered. Gynecological tumor markers did not show any obvious abnormalities. When patient underwent surgery for treatment, a mass was found at the root of suspensory ligament of the ovary. After resection and removal of the mass laparoscopically, post operative pathology and immunohistochemistry suggested Anastomosing Hemangioma.

Keywords: Anastomosing Hemangioma, Suspensory Ligament, Ovary

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INTRODUCTION:

Anastomosing hemangioma is a benign neoplasm of vascular origin similar to angiosarcoma. This tumor mostly consists of irregular anatomosing sinusoidal spaces lined by epithelial cells, where cells of the neoplasm are well differentiated and show very less atypia. Usually this tumor has defined boundaries with minimal invasion.[1] This lesion was first described and termed by Montgomery and Epstein in 2009.[2] These tumors are usually restricted to the Genitourinary tract but other locations have been reported like liver, adrenal glands, gastrointestial etc.[3] Here, we discuss an unusual presentation of such a tumor at the root of suspensory ligament of the ovary. Suspensory ligament of the ovary attaches ovary to the pelvic wall it is also known as infundibulopelvic ligament. Clinical significance of this ligament is it holds the ovarian blood vessels, nerves and lymphatics. Anatomically it is directed upwards towards the iliac vessels.

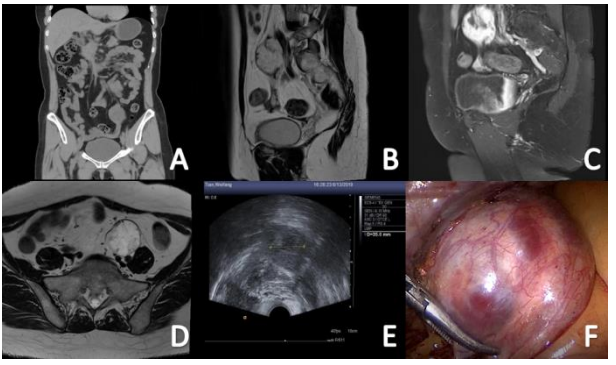
CASE REPORT:

A female patient 53 years old, who had natural menopause 3 years ago presented with an incidental pelvic mass on physical examination about a month before arriving at our hospital. The patient was hospitalized for further investigation and treatment. At the time of admission patient did not present with any complains of abdominal pain or back pain.

CT scan showed a slightly higher density shadow, a mass, about 36.5x35.0 mm infront of the psoas major muscle in the pelvis (fig.A). MRI examination of showed a left iliac paravascular T1 and long T2 signal shadow about 36.8x45.0 mm in size, mass showed progressive filling to the center (fig. B, C,D). this suggested the mass could be either schwannoma or tumor of adnexal origin possibly a sex cord stromal tumor. A gynecological ultrasound of the left adnexa shows 42x41x35 mm heterogeneous, low echo mass of irregular shape , unclear boundaries, poor echo with no blood flow and a 2.4x4 mm dark cystic area is seen inside the mass (fig.E). Gynecological tumor markers had no obvious abnormalities. Three days after admission patient received a laparoscopic surgery for further investigation and

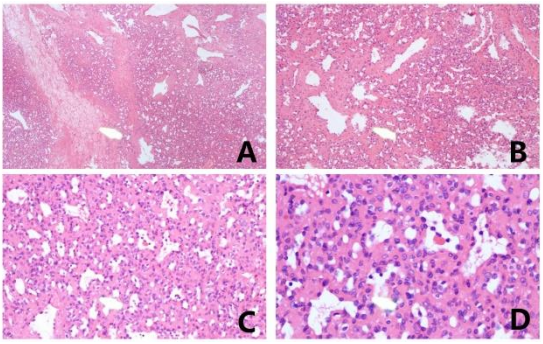
possible resection of the mass. Surgery revealed a tumor about 4 cm in diameter at the root of suspensory ligament of the ovary with dense adhesion(fig.F). After clearing the adhesion, laparoscopic left salpingo-oophorectomy along with left suspensory ligament tumor removal was carried out. Post-operative pathological exam of the left adnexa and the tumor showed: chronic inflammation of fallopian tube, corpus albicans was seen in the ovary and tumor pathology suggested possibility of anatomosing hemangioma which was to be further confirmed by immunohistochemistry. On immunohistochemistry, the cells of the tumor mass were CD31 (+), CD 34(+), ERG (+), CD56(+), NSE(-), Inibin A (-), D2-40(-), Ki67(5%+). these results combined with the HE stained sections on histology(fig.G) and clinical findings (i.e. clear boundaries of the tumor) are all consistent with anastomosing hemangioma. The patient had a smooth post-operative recovery without any complications. She was discharged on the 5th day post-operatively without any follow-up treatment due to the benign nature of anastomosing hemangiomas. Patient had no further complaints or discomfort during the follow-up checks.

Figure 1:



A- CT scan shows a shadow in the left pelvic cavity in front of the psoas muscle.
B- MRI a mass can be seen in near the left iliac vessels.
C- MRI enhancement shows a high density shadow in the left pelvic cavity.
D- MRI.
E- Transvaginal Ultrasound shows mass with poor echo, no blood flow and central cystic area.
F- 4cm mass found on laparoscopic surgery at the root of suspensory ligament of the ovary, mass has blear boundaries with dense adhesion.

Figure 2:



Microscopic appearance: capillary like architecture with irregular sinusoidal spaces lined by endothelial cells. (A: H&E, 40×; B: H&E, 100×; C: H&E, 200×; D: H&E, 400×).

DISCUSSION:

Anastomosing hemangioma (AH) is a subtype of capillary hemangioma, earlier thought to occur only in the genitourinary tract especially kidney, but other sites also include area next to the spine, adrenal glad, liver, stomach and ovary.[3,4,5,7] The lesion is usually benign in nature and pathologically has been defined as predominantly containing irregular anastomosing-sinusidal spaces with epithelial cells at the margins and very less atypia. Borders of the tumor seldom invade the surroundings and has a hobnail appearance.[1] Montgomery et al.were first to describe this lesion in 2009, they reported 6 cases which occurred in kidney and the lesion pathologically resembled the red pulp of the spleen.[2,8] AH itself is a less common tumor type in which ovarian AH are very rare with very few mentions in literature.[3] Though in most cases of ovarian AH the gynecological tumor markers are within normal range literature reports one case with massive ascites and elevated CA125.[6] AH has non-specific image findings on radiology, it is usually misdiagnosed as other rapidly progressive neoplasms or highly vascular tumors.[1,4,5] Both CT and MRI have no characteristic presentation when it comes to AH. Unenhanced axial CT scan shows a round mass with well-circumscribed borders, appearing heterogeneous. MRI often shows a space occupying lesion with clear boundaries, peripheral enhancement and uneven intermediate density. It is difficult to make a definitive diagnosis based on imaging alone.[7] Preoperative diagnosis of this

tumor is not possible, a definitive diagnosis is based on pathological and immunohistochemical exam of the tumor tissue. Histopathology of the tumor is constantly described as tightly packed anastomosing capillary-sized vessels with spaces resembling splenic sinusoids, the tufts of capillaries are lined by endothelial cells with absent or minimal atypia.[1,3-5,7-9]

On high magnification the hobnail endothelial cells and fibrous mesenchymal cells form a web like architecture.[2,10] Immunohistochemical exam shows sparse staining for endothelial markers like CD31, CD34, factor VIII, ERG, and FLI1.[1-5,7-9] Ki-67 (low poliferation of endothelial cells), GLUT-1(juvenile hemangioma), CD8(splenic sinusoids), and D2-40(lymphatc origin) immunostains are usually negative. Epitelial markers, Kaposi sarcoma antigens and Human Herpes Virus 8 antigens, S-100 protein, CD 117, HMB45, human placenta alkaline phosphatase and hCG all show no immunoreactivity when it comes to AH endothelial cells.[1]

In our case the patient's condition was insidious and lacked any specific clinical features. Patient had an incidental finding of a pelvic mass on physical examination. On imaging studies, CT scan showed a slightly high density shadow about 36.5x35.0mm in size infront of the left psoas major muscle and MRI showed TI signal shadow on the left side of iliac blood vessels of about 36.8x45mm in size with progressive filling to the center. Patient's gynecological tumor markers were within normal limits. Depending on the location of the mass and clinical findings possibility of a shwannoma or sex cord stromal tumor was considered. This lesion is rare, difficult to diagnose based on imaging and difficult to distinguish whether benign or malignant pre-operatively.

It is important to differentiate this tumor from schwannomas, sex cord tumors and most importantly from angiosarcoma. Schwannomas are benign tumors that usually arise around peripheral or cranial nerves. Common site for such lesion being the base of the tongue. They are well demarcated, minimally invasive lesions which are usually diagnosed using MRI and confirmed by S-100 protein positive on immunohistochemistry. Tumors are usually encapsulated unlike AH which usually lack capsule.[11] Sex cord stromal tumors of the ovary are also a rare entity which affects a broad age group. They can be classified into pure sex cord tumors, pure stromal tumors or

mixed type. They can arise from single cells like granulosa cell tumor or theca cell tumor. They can also arise from combination of different cells such as granulos-theca cell tumor or sertoli-leydig cell tumor. Many types of sex cord stromal tumors are associated with clinical syndromes, endocrine disorders and abnormal hormonal levels. Diagnosis is mainly based on the tumor's unique pathological morphology.[12] Angiosarcoma is a rare soft tissue tumor which is malignant in nature. Angiosarcoma of the ovary is very rare and its clinical features are non-specific mainly including abdominal pain, abdominal or pelvic mass and elevated CA125.[13] Microscopically, the appearance of the tumor tissue is complex and comprised of endothelial cells of various degrees of differentiation. It can form lumen when differentiation is high and tube like structures on low differentiation. Only a single cavity or microcapsule is seen.[13,14] Though AH resembles angiosarcoma, characteristic sinusoidal pattern, minimal differentiation of endothelial cells and the absence of epithelial neoplastic cells in an AH should assist in differentiating the two lesions.[1]

AH is a neoplasm of benign nature hence surgical resection of tumor is preferred option for treatment and it has fairly good prognosis.[15,16]

CONCLUSION:

According to majority case reports in literature AH is a benign, slow growing lesion with good prognosis when treated with surgical resection, which occurs not only in genitourinary tract but also at various other locations in the body. There is no recurrence, metastasis or death from the disease after follow-up. The diagnosis in this case is mainly based on pathological and immunohistochemical exam. Misdiagnosis of the lesion pre-operatively is likely due to less specific clinical characteristics or non specific radiological findings. It should be considered as differential in rare type pelvic masses. It is important to distinguish this tumor from similarly presenting rapidly progressive or highly vascular lesions like angiosarcoma to prevent over treatment. The characteristic presence of sinusoidal pattern and anastomosing capillaries with no or less atypia of enodthelial cells on

histology with positive CD31 and CD34 on immunohistochemistry should help differentiate this lesion.

Declaration of Patient Consent:
Authors confirm that the patient was informed that their clinical information and subsequent imaging results might be published in the journal and the relevant informed consent forms were signed by the patient. The patient is aware that his/her name and identity will not be published and concealed to the best of our ability. The patient understands that complete anonymity cannot be guaranteed.

Conflicts of Interest:
None.

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