FINAL DIAGNOSIS: Angioleiomyoma (AL) of uterus

DISCUSSION

Leiomyomas are the most common uterine neoplasms. There are different variants of leiomyomas and amongst these variants AL is extremely rare as till date only fifteen cases of AL of the uterine corpus have been described in the English literature and one case in the Korean literature.[1-13] Uterine AL usually occurs in the middle aged females. The age range in the cases described is from 30 years to 69 years. Most commonly the patients present with menorrhagia, pain abdomen and/or abdominal mass. Marci et al have described a case of consumptive coagulopathy secondary to a large degenerated AL.[1] Rarely the patient may present with Pseudo-Meigs syndrome and raised CA125 levels, thereby mimicking an ovarian neoplasm.[2] Single case of AL in a patient with tuberous sclerosis has also been described by Jameson et al.[11]

AL commonly present as a well-circumscribed mass arising from the uterine corpus. Few cases with multiple uterine AL have also been reported.[1,8] They range from 4cm to 26cm in the maximum dimension.[1-12] These can be submucosal, intramural, and/or subserosal.[1-12] Those arising in the posterior myometrium in a subserosal location may present as a pelvic mass and may be difficult to differentiate from primary ovarian neoplasm preoperatively.[3] On sectioning, the cut surfaces of AL are tan-white, whorled with hemorrhagic areas, or may exhibit a variegated appearance with pink-brown and gray areas.[8] Some are multiloculated with blood filled cysts.[7] Histologically, AL is composed of interlacing fascicles of spindle cells with interspersed abundant thick walled blood vessels. The monotonous and spindled smooth muscle cells swirl around the blood vessels. This is in contrast to the usual leiomyomas in which the density of the vascular network is similar to or less than the normal myometrium.[19] Moreover, the blood vessels in the usual leiomyoma are predominantly capillaries along with a few arterioles and small arteries in contrast to the thick walled muscular vessels as seen in AL. Most of these lesions lack mitotic figures, pleomorphism, or necrosis.

However, Thomas et al observed marked atypia with bizarre hyperchromatic nuclei and multinucleated giant cells in their case.[2] Focal nuclear atypia was present in another case described by Sahu et al.[3] However extensive sampling in both the abovementioned cases revealed no excess or atypical mitotic activity, coagulative necrosis, or hypercellularity. Mitosis up to 2 mitoses per 10 high power fields was seen in one case.[5] All these features emphasize the importance of extensive sampling in cases where there is nuclear atypia or mitosis in order to exclude a leiomyosarcoma. Other changes that have been described in this tumor are myxoid change, edema and hyalinization of the stroma,[2,3,5,8] and fibrin deposition in the vessel wall.[5] Even though fibrin thrombi within the vessels are characteristically present in AL of the skin,[20] only two