Angiomyolipoma in the Breast and Review of Literature

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Angiomyolipoma (AML) is a hamartomatous disorder composed of varying proportions of smooth muscle, adipose tissue, and blood vessels, with the kidney being the most frequent site of involvement. Many cases are associated with tuberous sclerosis; however, majorities of patients do not have this disease complex (1). They have also been encountered in extrarenal sites such as the liver (2), lungs (3), penis (4), fallopian tube (5), colon (6), spinal cord (7), skin (8), hard palate (9), intramyocardium (10) and spermatic cord (11). We review the literature and offering the breast AML first. Its treatment is the total resection of the mass. In most organs, it may give rise to misdiagnosis in biopsy especially with fine-needle aspiration biopsy (12).

A thirty-four-year old woman, upon noticing a growth enlarging slowly in her left breast, presented with a painless mass complaint. Her history revealed that seven years previously she had had 2x2 cm cystic higroma excised in the right elbow, and 5 years previously three lipomas in the left wrist and right lower extremity, the biggest being 3 cm in diameter. There was no tuberous sclerosis. CT and US revealed heterogeneous echoic masses in the upper outer quadrant of the left breast. The findings from biochemical analyses were within normal limits. Receiving the preliminary diagnosis of the breast lipoma, the patient underwent breast salvage surgery in which the breast was totally removed with its capsule. One week after the surgery the patient was discharged with complete recovery. The removed mass was a capsuled light yellow to tan one-size 12x8x6 cm with medium consistency (Figure 1).

Histopathologically, the tumour consisted of a mixture of mature adipose cells, thick-walled blood vessels, and spindle smooth muscle cells. The spindle smooth muscle cells appeared to have originated from the walls of the medium sized and large thick-walled vessels, and proliferated centrifugally in the form of sweeping fascicles, sheets or isolated cells among mature adipose cells (Figure 2). The proliferated spindle smooth muscle cells were eosinophilic cytoplasm and, their nuclei elongated and were vesicular. Cells with bizarre nuclei were not common.

The spindle smooth muscle cells yielded positive reactivity with desmin, vimentin, and smooth muscle actin, and HMB-45 (Zymed). S-100 protein was positive in adipocytes only, whereas cytokeratin and epithelial membrane antigen were negative.

AML is a benign tumour considered hamartomatous. It occurs more frequently in women and its median age is 46. Most of the cases are asymptomatic, and therefore detected either incidentally or in biopsies performed for another reason (1). They can be solitary or multifocal, with the kidney being the favoured location. Forty percent to 80% of patients with tuberous sclerosis have renal angiomyolipomas, but approximately 80% of renal angiomyolipomas occur in patients without symptoms of the disease. In those occurring with tuberous sclerous complex, hyperpigmente pots, shagren patches, preunical fibromas, renal cysts, cardiac rhabdomyoma and calcification of the cerebral cortex with mental deficiency may be seen (1). Lymphangiomyoma, renal carcinoma, and neurofibromatosis have been reported rarely (3). Extra-renal AML, the liver (2), the lung (3), several cases of spermatic cord involvement (11), two cases with subgaleal involvement (13), one case with nasal cavity involvement (14), and other sites have been reported.
Macroscopically, its size may range from several cm to 20 cm. In our patient, it was 12 cm in diameter. Haemorrhage, focal necrosis, cystic degeneration and focal dystrophic calcifications are frequent and these variations may present difficulty in differentiating it from malignant tumour (1). Microscopically it is composed of blood vessels, smooth muscle and mature adipose tissue. One of its components may be less and thus wrongly may receive such diagnosis as angiolipoma, vascular leiomyoma (often painful), and myolipoma when sampling was inadequate (1). Hamartomas present either as a palpable mass or as discrete lesions detected mammographically. Radiographically, the lesion appears as a well-defined density, surrounded by a narrow zone of radiolucency and often gives the appearance of being encapsulated. They range in size from 1 to 13.5 cm, with a mean of about 3 cm (15). Since atypical spindle cells and lipoblastic cells in fine needle aspiration cytology may give rise to misdiagnosis, using smooth muscle cell markers immunohistochemically will help approach the diagnosis (12). One interesting and rare aspect of this tumour is that it may be seen together multicentrically in the kidney and lymph node (16). It occurrence in the lymph node is not considered metastasis.
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de Jong et al. reported trisomy 7 in a patient with angiomyolipoma (17). In a study of DNA content analysis by Abdulla et al., all of the six multicentric AML cases were aneuploid pattern, while aneuploid pattern was detected in one of the four cases with classic renal AML (18). Interestingly diploid pattern was detected in a patient with hepatic, splenic and pulmonary lesions. The presence of aneuploid pattern is not reliable marker of malignancy in these cases. In a study Abdulla et al., immunohistochemical analysis revealed positive staining reaction of vascular and adipose tissue components with HMB-45 antibody in three of the six cases of AML (18). Few immunohistochemical investigations on AML have been published. These studies have emphasized its reactivity for desmin and vimentin. The surprising finding of immunoreactivity of AML with HMB-45, an antibody that melanocytes and is thus an important marker of malignant melanoma, has been recently reported (18). Reactivity for HMB-45 of the polygonal smooth muscle cells was first reported by Pea et al. (19). In contrast to the findings of Pea et al. (19), a study by Chan et al. (20) shows that HMB-45 reactivity occurs not only in epithelioid cells, but also frequently in the spindle cells. AML positivity rates for smooth muscle actin, desmin, and HMB-45 were: 20/20, 17/20 and 18/20 respectively, including one case that was negative for both desmin and HMB-45, as in the study by Chan et al. (20).

Hoon et al. (21) and Peat et al. (19) have grouped these reactive cells under the heading perivascular smooth-muscle cell. Bonetti and colleagues state that the histogenesis of the perivascular smooth-muscle cell is uncertain but that the close relationship of these cells with the vessel wall is suggestive of their origin from this structure (22). The validity of at least a portion of this theory has to be questioned.

Contrary to the fact that angiolipoma, lipoma, myelolipoma, and vascular leiomyoma are frequently observed in the breast, we were not able to detect any occurrence of AML in the relevant literature (1, 15). Research is needed on a large series of cases in order to be able to understand the histogenesis, nature, and malignancy potential of AML cases.

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