Cytological Diagnosis of Mazabraud’s Syndrome

Sir,

Mazabraud’s syndrome is characterized by a combination of benign intramuscular myxomas and fibrous dysplasia of bone. It was first described by Henschen in 1926 and Mazabraud in 1957. Both myxoma and fibrous dysplasia are benign entities but their occurrence in combination is quite rare with around 81 cases reported till date. Here, we report one of the very few cases of Mazabraud syndrome in a male diagnosed on cytology.

CASE

A 20-year-old male presented with a painful gait and slow mass growing in his right calf for 3 years. The mass was 6 × 4 × 3 cm in size and was firm and nontender. X-ray of the right tibia and ipsilateral femur showed similar lesions with characteristic ground glass appearance suggestive of fibrous dysplasia. Further, X-ray of the skull showed an osteoma.

Fine needle aspiration of the calf mass yielded clear, gel-like material and the tibial lesion yielded blood. Smears prepared from the gelatinous material showed few spindle cells in a myxoid background. These spindle cells had oval-to-spindled nuclei with bland chromatin along with long cytoplasmic processes that were intertwined at areas. Few thin-walled capillaries were also noted [Figure 1a]. Cytological diagnosis of an intramuscular myxoma was made.

Smears from the tibial lesion yielded only blood along with a few scattered bland appearing spindle cells. Definite opinion was not possible and biopsy was advised.

Histology of biopsies from the tibial and femoral lesions showed curvilinear trabeculae of metaplastic woven bone in hypocellular and fibroblastic stroma without osteoblastic rimming; a diagnosis of fibrous dysplasia was made.

The combined radiological, cytological, and histopathological findings combined were consistent with the diagnosis of Mazabraud’s syndrome.

DISCUSSION

The combination of polyostotic fibrous dysplasia and intramuscular myxoma has been called Mazabraud syndrome. Most cases present with polyostotic fibrous dysplasia, however, mono-ostotic lesions have been described as well. Myxomas appear in adulthood while the bony lesions start to occur in childhood. It is twice as more common in females than in males.

It was also noted that most cases of this syndrome involved the lower extremities and that there can be predilection to one side of the body.

Moreover, the benign, symptom-free myxomas usually develop in the muscle next to the most extensively involved bone with fibrous dysplasia.

Until 2012, only 81 cases of Mazabraud’s syndrome have been reported. Although the prognosis of most fibrous dysplasia are good, the polyostotic fibrous dysplasia associated with Mazabraud’s syndrome has a higher risk of malignant transformation and thereby mandates appropriate and early diagnosis for constant monitoring. Malignant transformation of myxoma has not been reported till date.

CONCLUSION

This case documents the cytomorphology of myxoma and emphasizes the need to be aware of Mazabraud syndrome, which will enable constant monitoring and early diagnosis of malignant transformation in such patients.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have...
given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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