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ABSTRACT

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CASE REPORT

A 45-year-old male presented with a swelling in the perianal area that had begun to grow four months previously. He complained of pain, and problems defecating. Physical examination located a soft lump in the perianal area, with ill-defined edges near to the posterior midline (Figure 1). Although it is a rare condition, one of our preliminary differential diagnoses was lipomas. Other differential diagnosis was possible Desmoids Type Tumor.

Magnetic resonance imaging (MRI) scan of the pelvis showed a large flask shaped mass sagging from the perineum, with fat lobules apparently originating from perineum, extending into the intergluteal cleft. There was concern about the swirled appearance noted post contrast study, which suggested possible diagnosis of a myxoid type tumor and less likely a lipoma (Figure 2A–B).

Surgical excision of the mass was performed. The gross appearance of the specimen was a disoriented mushroom shaped mass, about 380 grams in weight, with a fatty

cut surface (Figure 3). Microscopically, it showed an ill-defined tumor composed of small spindle cells embedded in an abundant myxoid stroma interspersed with a thick wall and hyalinized blood vessels. There was no evidence of mitotic activity or nuclear atypia (Figure 4A–C). Based on these histological features, a diagnosis of aggressive angiomyxoma (AAM) was confirmed.

DISCUSSION

Aggressive angiomyxoma (AAM) is a rare benign soft tissue tumor of mesenchymal origin, characterized by its

Figure 1: Gross appearance of soft lump in the perianal area, adjacent to posterior midline with ill-defined edges and about 28 x 22 cm.

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Received: 20 February 2015

Accepted: 21 March 2015

Published: 01 July 2015

Figure 2: Magnetic resonance imaging (MRI) scan of pelvis (A) Coronal section T1 W image showing Large Flask shaped mass sagging from Perineum and measures 29 x 22 x 17 cm. MRI of pelvis, (B) Sagittal Section image T2 W image : Swirls of T2 hypointensity is noted within the Mass which is characteristic of AAM (arrow) .

Figure 3: Gross appearance of the specimen showing Disoriented mushroom shaped mass, and about 380 grams in weight, with a fatty cut surface.

Figure 4: (A) An ill-defined tumor composed of oval to spindle shaped cells lacking nuclear atypia with no mitotic activity, (B) A characteristic feature of aggressive angiomyxoma is the appearance of lesional cells spinning off a vessel that they encircle (arrow), (C) Note the thick-walled larger vessels (H&E stain, x100).

locally infiltrative nature and high recurrence rate. It was given this name due to the pathological findings of the stellate and spindled cells, along with variable-sized blood vessels, which were intricately entwined within a myxoid matrix [1]. This presentation occurs predominantly in women of reproductive age (female to male ratio of approximately 6:1), and exclusively in the pelvi-perianal region. To our knowledge, only 43 cases occurring in men have been reported in literature [1]. In men, AAM usually involves the scrotum (38%), spermatic cord (33%), and perineal region (13%) [2].

Although clinical diagnosis of AAM may be difficult, the typical MRI features of AAM are swirled strands, aligned with the craniocaudal axis. This particular imaging feature is caused by a stretching of the fibrovascular stroma [3]. Confirmation of diagnosis is based on a histopathology study. AAM should be distinguished from other benign, potential lesions with low local recurrence, and malignant tumors with widespread metastatic potential.

Surgery is the mainstay of treatment for AAM, although achieving negative resection margins is complicated, because of the infiltrative nature of the tumor. Local recurrence rate falls between 36% and 72% and is usually seen in the first three years. Recurrences of the disease are usually controlled with surgery [1]. Several reported attempts at using chemotherapy and radiotherapy as part of the treatment protocol for AAM have proved disappointing; probably due to the low mitotic activity/growth fraction of cells. Primary treatment with GnRH agonists has proved successful, as many of angiomyxoma are positive for estrogen and progesterone receptors [4]. However, the duration of the responses and an optimal treatment schedule are still unknown [1].

CONCLUSION

In conclusion, whether the treatment of aggressive angiomyxoma (AAM) is surgery, hormonal therapy or a combination of the two, it is apparent that adequate management of AAM necessitates close, and long-term follow-up at sixth month intervals, to monitor for recurrence. Multiple relapses can occur but metastases are unusual.

How to cite this article

ALHumoud ZT, Aldaoud N, Abrar H, Salem A. Perianal aggressive angiomyxoma in a male patient. Int J Case Rep Images 2015;6(7):454–456.

doi:10.5348/ijcri-201522-CL-10077

Acknowledgements

We would like to thank Dr. Thabet Ghazal who was involved in the Management of the case.

Author Contributions

Zainab Taha ALHumoud – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Najla Aldaoud – Substantial contributions to conception and design, Drafting the article, Final approval of the version to be published

Hussain Abrar – Substantial contributions to conception and design, Drafting the article, Final approval of the version to be published

Amro Salem – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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SUGGESTED READING

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