A CASE OF RARE DESMOID-TYPE FIBROMATOSIS PRESENTED AS A BREAST LUMP

Wan Irfan Bin W Mustapha*[1]; Nur Shahida Binti Wahab[2]; Radhiana Binti Hassan[3]; Raihanah Binti Haroon[4]
[1-4]*Radiology/ International Islamic University Malaysia/ MALAYSIA*

INTRODUCTION

Desmoid-type fibromatosis (DF) is a benign but rare soft tissue tumour, occurring only in 2-4 persons in one million of the population . Its local invasiveness and high tendency to recur following resection contribute to its notorious reputation for being an aggressive type of fibromatosis. It may be encountered across all demographic but more commonly seen between the second and fourth decade of life with a female predilection. As it originated from the musculoaponeurotic structures throughout the body, it may affect any site, with the limbs and abdomen being the more commonly reported encountered sites. Chest wall DF only represents about 8-10% of the incidence. In this report, we describe our encounter with a case of chest wall DF and how it presents in the different radiological modalities that we have in our centre.

DISCUSSION

Desmoid-type fibromatosis is known to have a higher prevalence involving the abdominal wall, with the extra-abdominal organs involvement discovered later. Chest wall DF is thought to account for 10-20% of all the cases. As there is only a fraction of chest wall DF, which has been reported in the literature, we believe the identification of its features in the various imaging modalities would be of paramount importance for the identification of such a rare case. Among the cases of extra-abdominal DF (also known as aggressive fibromatosis), they are often described as a slow-growing tumour without metastasis but with a high risk of local invasion and local recurrence following excision. Our patient presented with a palpable mass on the right 'breast'. The initial clinical examination was difficult as the patient was thin with relatively small breasts volume, rendering the breast tissue indiscernible from the underlying chest wall. The mammogram revealed a dense mass that was partially seen on the mediolateral oblique view. No additional information was obtained from the tomosynthesis; however, we were able to identify a large heterogeneous lesion in the lateral aspect of the right breast measuring 3 x 6 x 2 cm with poor visualization of its relation to the posterior structures. On the CT, the lateral aspect of the right pectoralis muscle was markedly thickened. It was gently displacing the fibroglandular tissue of the right breast anteriorly, but no focal lesion was seen within the muscle or the right breast. Following this, an MRI breast was performed. In this imaging, we identified an irregular lobulated mass measuring 1.7 x 4.5 x 2.4cm (AP x W x CC) at the right upper outer quadrant of the chest wall with a rounded non-enhancing region at the medial edge of the mass measuring about 1.3 x 1.3 cm likely to represent cystic or necrotic content. The mass was isointense to the muscle on T1W, slightly hyperintense on T2W with contrast enhancement on post gadolinium. No area of restricted diffusion noted on echoplanar imaging. It was continuous with the right pectoralis major muscle, with the expansion of the right pectoralis muscle laterally. Anteriorly the mass was causing displacement of the right breast tissue anterior and medially. There were areas of the diminished fat plane between the mass and the breast tissue, likely to represents tumour infiltration from the pectoralis muscle into the adjacent breast tissue. Posterior and inferiorly, there was also an invasion of the right external intercostal muscle without any intrathoracic extension. Following the findings of a deep intramuscular/soft tissue mass origin with local structural invasion, we believed that a malignant soft tissue tumour (sarcoma) was most likely. However, the mastectomy sample yielded a result of an ill-defined tumour composed of spindle cells arranged in long sweeping fascicles with some vague storiform pattern without any significant atypia, which was concluded as desmoid-type fibromatosis. The nearest margin was 1 mm (deep margin) and on immunohistochemistry expressed vimentin, beta-catenin mutation, smooth muscle actin and PS100.

"I have no potential conflicts of interest to report with regards to this presentation".

CASE

A 46-year-old lady presented to the Department of General Surgery Outpatient Clinic of IIUM Medical Center with a complaint of a right breast lump which had been present since the past few years but felt a sudden increase in size prior to her presentation. She had no associated symptoms or family history of malignancy. Clinical examination revealed a 4 x 5 cm hard mass at 9 o'clock in the right breast, which was fixed to the underlying pectoralis muscle. No other stigma of breast disease was identified. A bilateral mammogram and tomosynthesis with complimentary ultrasound breasts were performed. Although mammogram and complementary ultrasound were able to identify a dense mass in the right breast, we were not satisfied with the posterior margin of the mass; therefore, a CT Thorax, Abdomen and Pelvis was performed to delineate the posterior structure and explore whether there is any distant metastasis. Following this, a trucut biopsy was taken, which revealed a mesenchymal lesion. Finally, magnetic resonance imaging (MRI) of the breasts was performed to better characterize the mass. The features of the mass on this imaging are discussed in the following section. This patient eventually underwent right mastectomy as a result of overall features of Desmoid-type fibromatosis.

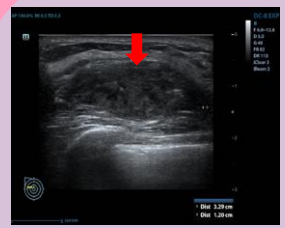

Figure 1: Transverse ultrasound image shows a large suspicious lesion is seen occupying the lateral part of the right breast (red arrow). It is situated adjacent to the areolar region and extends toward the right axilla. It measures about 3.3 x 1.2 cm. Some part of the lesion shows ill-defined margin. Unable to delineate posterior margin and involvement of the pectoralis muscle. The lesion also shows heterogeneous echogenicity. No increase in surrounding vascularity.

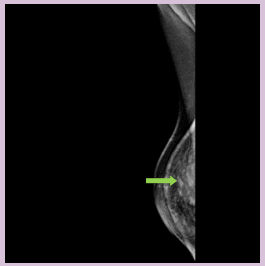

Figure 2: Right breast is dense (BIRADSD). There is a mass that is partially seen on the MLO view (green arrow). The lesion measures about 3 cm. No obvious stromal distortion seen. No suspicious clustered microcalcification. Slight skin thickening is seen, but no nipple retraction. Tomosynthesis images do no add value in this case.

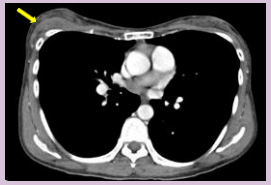

Figure 3: The lateral aspect of the right pectoralis muscle is markedly thickened with maximal thickness measuring about 1.3 cm (yellow arrow). This is seen gently displacing the fibroglandular tissue of the right breast anteriorly. Adjacent subcutaneous tissue stranding is seen. However, no focal enhancing mass lesion is detected within the muscle thickening, which appears isodense to surrounding muscle or the fibroglandular tissue.

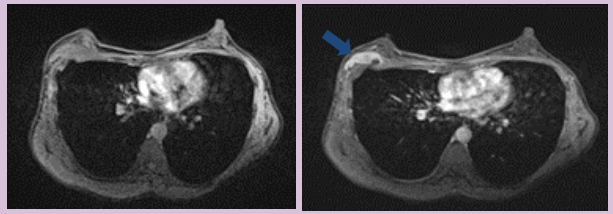

Figure 4: The mass is vividly enhanced on post gadolinium (blue arrow). It is continuous with the right pectoralis major muscle, with an expansion of the right pectoralis muscle laterally. The mass is causing displacement of the right breast tissue anteromedially.

CONCLUSION

As extra-abdominal fibromatosis is rare, its history of slow growth, the depth in the location of its origin and local invasiveness on multimodality imaging should be a foundation for its diagnosis with the aid of immunohistochemistry staining on pathology.

REFERENCES

moorthy Ganeshan, D., Amini, B., Nikolaidis, P., Assing, M., & Vikram, R. (2019). Current update on desmoid fibromatosis. *Journal of computer assisted tomography*, *43*(1), 29.
Kabiri, E. H., Al Aziz, S., El Maslout, A., & Benosman, A. (2001). Desmoid tumors of the chest wall. *European journal of cardio-thoracic surgery*, *19*(5), 580-583.
Zehani-Kassar, A., Ayadi-Kaddour, A., Marghli, A., Ridene, I., Daghfous, H., Kilani, T., & El Mezni, F. (2011). Desmoid-type chest wall fibromatosis. A six cases series. *Orthopaedics & Traumatology: Surgery & Research*, *97*(1), 102-107.