Intraosseous hibernoma: A case report and review of the literature

Hibernoma intraosseux: A propos d'un cas et revue de littérature

Manel Yahia, Besma Laabidi, Issam M'sakni, Fethi Bougrine, Ammar Bouziani

Service d'Anatomie et de Cytologie Pathologiques-Hôpital Militaire de Tunis / Faculté de Médecine de Tunis

RÉSUMÉ

L' hibernome intraosseux est une tumeur osseuse bénigne rare, avec seulement 4 cas rapportés dans la littérature anglaise.

Observation : Il s'agissait d'un homme âgé de 42 ans sans antécédents pathologiques qui a consulté pour des douleurs thoraciques droites. L'imagerie a montré une lésion ostéolytique à composante graisseuse de la quatrième cote droite. La lésion a été complètement réséquée et l'étude microscopique a conclu à un hibernome intra-osseux.

Conclusion: Bien qu'il soit rare, l' hibernome doit être inclus dans le diagnostic différentiel de toutes les tumeurs adipocytaires.

Mots-clés

SUMMARY

Intraosseous hibernoma is a rare benign bone tumor, with only 4 cases reported in English literature.

Case report: In this report, we describe a 42-year-old man with no past medical history and right chest pain. Imaging studies showed an osteolytic lesion with fat attenuation lesion in the right fourth rib. The lesion was completely resected and microscopic study showed mildly thickened bone trabeculae and multivacuolated brown fat cells replacing the normal white fat and hematopoietic elements. The diagnostic of intraosseous hibernoma was made.

Conclusion: Although it is rare, hibernoma should be included in the differential diagnosis of lipomatous tumors.

Key-words

Hibernoma, chest, tumors

Hibernomas are uncommon benign soft tissue tumours mimicking brown fat. This tumor was first described as pseudolipoma by Dr H Merkel in 1906 [1]. The most common anatomic locations include the neck, axilla, mediastinum, periaortic and perirenal zones [1]. Intraosseous and in particular costal locations are exceptional, there were only 4 cases reported [1]. Using a case report, we will discuss the diagnostic features of hibernomas and review the literature.

CASE REPORT

A 42-year-old man with no past medical history, was admitted for a 6-month history of a right chest pain without fever, chills, night sweats or weight loss. Physical examination was normal. Chest radiograph showed a low density mass in the posterior arch of the right fourth rib. Computed tomography (CT) scan showed an expansive and lytic bone lesion of the posterior arch of the right fourth rib, without periosteal reaction or cortical rupture. The mass showed a fat attenuation (fig.1).

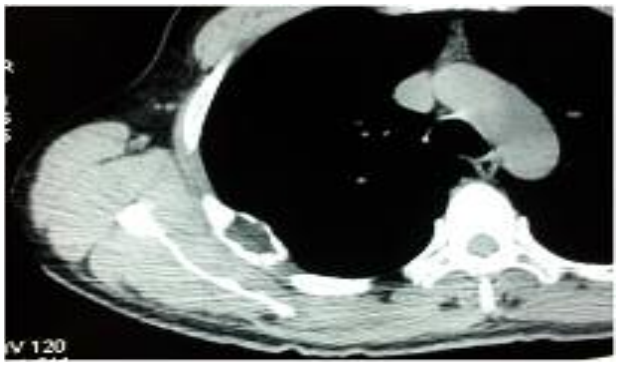

Figure 1 : CT scan of the thorax: osteolytic and sharply demarcated mass arising from the posterior arch of the right fourth rib. Fat attenuation was observed wich suggested the adipose nature of the tumor.

A bone scintigraphy was performed showing an uptake of the posterior arch of the right fourth rib without osteolytic lesions. As malignancy could not be excluded with certainty, the decision was made to completely resect the lesion. The chest was entered through a posterolateral thoracotomy. A large tumour with a smooth and gleaming capsule was visualised. A resection of the posterior arch of the right fourth rib was performed. Gross examination of the rib showed a mass tan to light brown color separated by fine fibrous trabeculae with lobulated, well-demarcated margins, and measured about 5 cm in size. Microscopically, numerous lobules separated by septa of connective tissue were present. Tumoral cell had granular and deeply eosinophilic cytoplasm (fig.2, 3). The nuclei were always small, regular and round. There was no atypia and no mitoses. The tumour had a thick intact capsule consisting of collagenous connective tissue and doesn't infiltrate the bone tissue and the cortical. The adjacent extra osseous tissue was free of tumor. The S 100 protein was positive (fig.4). The CD34 was negative. The diagnosis of hibernoma was made. Clinical examination and chest radiograph after 6 months were normal.

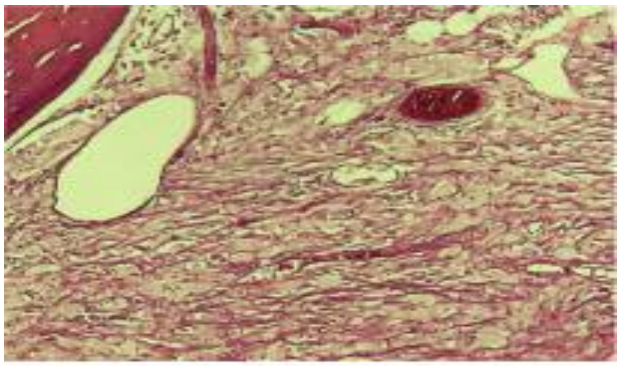

Figure 2 : (HEX 200): Granular, multivacuolated, eosinophilic, round to oval cells with centrally placed nuclei and multiple small lipid droplets.

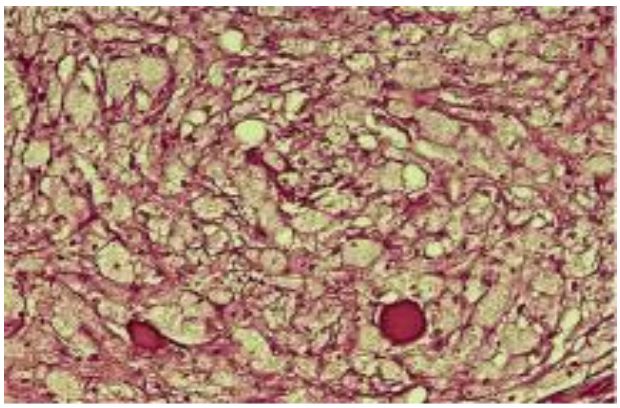

Figure 3 : (PAS X 400): weakly positive.

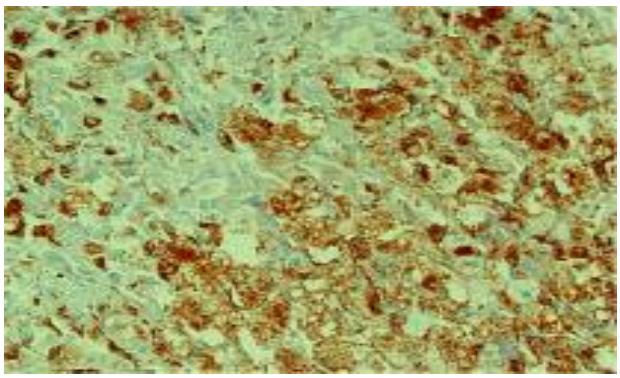

Figure 4 : S-100 protein stain (X400) was positive, which ruled out metastatic granular renal cell carcinoma

DISCUSSION

Hibernoma is a rare benign tumor consisting of brown fat [3]. Soft tissue hibernoma is rare, occurring most commonly in the thigh [3]. Our case is the fifth documented case of intraosseous hibernoma. Interestingly, all the cases of intraosseous hibernoma involved bones of the pelvis or lower extremity (table 1). Our case involved in the rib.

Table 1 : Clinicopathologic features of the 4 intraosseous hibernomas

Sex	Age (y)	Lesion site	Radiologic findings	Underlying condition	Source
F	57	Left sacral ala	Heterogeneously hyperintense with increased vascularity	Low back pain	Kumar et al [23]
M	54	Bilateral femur	N/A	N/A	Reyes et al [24]
F	60	N/A	N/A	Essential thrombocythemia	Thorns et al [25]
F	50	Right ilium	Sclerotic	Breast carcinoma	Bai et al

Abbreviations: F, female; M, male; N/A, not available.

Hibernomas are mostly seen in the fourth and fifth decades of life but the reported age ranges from 2 to 75 years. Previously a female predominance was reported, but in a large study of 170 cases published in 2001, 99 tumours occurred in male patients [9]. Our patient was 42 year old.

The most common site is the thigh, followed by the trunk, upper extremity and head and neck. The myxoid and spindle cell variants tend to be located in the posterior neck and the shoulders, similar to spindle cell lipoma. Less than 10% occurs in the abdomen or the chest.

Clinically, they are usually painless and are therefore often incidentally found during routine investigations. When symptoms are present, they often relate to compression of adjacent structures [11]. Significant weight loss is described and is attributed to excessive thermogenesis of the tumour tissue responsible for the catabolism of circulating lipids and carbohydrates into thermal energy [12]. Our patient presented right chest pain without fever or weight loss.

Hibernomas have CT and MRI appearances similar to other benign and malignant fibrous and lipomatous tumours [13]. The few recorded examples of hibernoma on CT [14] describe them as well circumscribed and heterogeneous lesions. The attenuation values range fat and muscle depending on the lipid content of the tumor and its vascularity, which is often prominent with branching serpentine small vessels [14]. There are also a few reported descriptions of MRI features of this tumor [14,18]. A recent study [6] highlights the value of MRI in differentiating simple lipomas (encapsulated homogeneous fatty mass with or without isolated thin septa) from lipoma-variants and well differentiated liposarcomas (heterogeneous fatty masses with nodular or thick septa, prominent foci of high T2 signal and prominent areas of enhancement after contrast administration). In the same article, the authors also point out the inability in distinguishing well-differentiated liposarcomas from lipoma-variants (where hibernoma is included). The differential diagnoses of a high T1 signal mass include angiolipoma, lipoma, hemangioma, liposarcoma, alveolar soft-part sarcoma, and clear cell sarcoma; however, none of them contains large intratumoral vessels typically seen in hibernomas [13]. Increased vascularity of a lesion could raise the suspicion of malignancy.

As needle biopsy carries a risk of haemorrhage and often leads to inconclusive results, definitive pathological diagnosis is based on surgical resection [20, 21].

Grossly, the tumors are well circumscribed, partially encapsulated, and lobulated. The cut surface varies from yellow to brown and is occasionally mucoid with rare areas of hemorrhage [3]. The diameter usually ranges from 5 cm to 10 cm, but it may reach up to 20 cm [4]. In our case, gross examination of the rib showed a mass tan to light brown color separated by fine fibrous trabeculae with lobulated, well-demarcated margins. The size of tumor was 5 cm. Microscopically, the presence of hibernoma cells,multivacuolated fat cells with small, central nuclei, was common to all tumors. Four histological variants have been identified based on the tinctorial quality of hibernoma cells, the nature of the stroma, and the presence of a spindle cell component: typical (82% of cases), myxoid (8%), lipoma-like (7%), and spindle cell (2%) [3]. Most tumours contain large numbers of multivacuolated brown fat cells with abundant, granular cytoplasm and a small, central nucleus, the granular or eosinophilic variant. The brown fat cells vary from pale staining to variably eosinophilic, and some cases have a mixture of pale and eosinophilic cells, the mixed variant, while other cases have pure pale brown fat cells, the pale variant. Some hibernomas contain small clusters of brown fat amidst ordinary white fat, the «lipoma-like» variant. Multivacuolated lipoblast-like cells are often seen. Rare variants with myxoid stroma (myxoid variant), or a spindle cell component, with thick bundles of collagen fibres, scattered mast cells, and mature adipose tissue (spindle cell variant), a hybrid between hibernoma and spindle cell lipoma, have been described. Mitoses are exceptional and cytological atypia is unusual. Such features should not be equated with malignancy. However, scattered normal brown fat cells may be found in an otherwise classic myxoid or well differentiated liposarcoma [22]. Hibernoma cells are variably, sometimes strongly, positive for S100 protein. The spindle cell variant has a CD34 positive spindle cell component, similar to spindle cell lipoma, whereas the other hibernoma variants are negative for CD34 [22]. In our

case the CD 34 was negative.
There is a critical role for pathologic diagnosis in these clinical circumstances. The morphological findings were similar to those usually described. The case favored the following diagnoses: hibernoma, metastatic granular renal cell carcinoma and granular cell tumor. For differential diagnosis, S-100 protein staining was positive, which ruled out metastatic granular renal cell carcinoma. Because periodic acid-Schiff staining was weakly positive and had no organoid pattern of the tumor, granular cell tumor was excluded.
Hibernoma is a benign tumour that does not recur with complete local excision. All morphologic variants have the same good prognosis [22].

CONCLUSION

Hibernoma is a rare benign tumor that arises from the vestiges of fetal brown fat. Although it is rare, hibernoma should be included in the differential diagnosis of lipomatous tumors. This is a benign tumor with no malignant potential. Complete excision is the treatment choice.

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