Angiomyxolipoma of the right sub-brow: Case report with review of the literature

Sami Al-Bdairat (M.D.)a,*, Isam Sh. Alrawi a, Mohammed A. Obaid a, Yahia F. Dajani b

a Eye Specialty Hospital, Amman, Jordan
b Grand Medical Laboratories, Amman, Jordan

1. Introduction

Lipoma is the most common benign mesenchymal tumor in adults, which may occur singly or multiple in any part of the body. Several variants of lipoma have been described and angiomyxolipoma is one rare subtype [2].

2. Case report

A 78-year-old lady attended the Department of Orbit & Oculoplasty in our hospital, complaining of a painless swelling on the right side of her forehead, involving the right sub-brow region. She noticed the lesion four months earlier with progressive increase in size. On physical examination, a mass 3.0 × 3.5 cm appeared on the right sub-brow region (Fig. 1). It felt elastic, firm and relatively mobile. The mass was non-tender, non-pulsatile and non-transilluminant. Overlying skin was normal. MRI with intra-venous contrast showed a cystic mass, hypo-intense on T1, hyper-intense on T2-weighted images, partially suppressed on FLAIR images and showed heterogeneous enhancement in post contrast images, MRI also showed very clearly the black-rim around the mass which represents the interface between the mass and normal adipose tissue. (Fig. 2). The lesion was surgically excised under general anesthesia, using a curvilinear incision over the mass itself. Intra-operatively, a cystic mass was showed a thin and delicate fibrous capsule that ruptured despite careful handling, with mucoid material issuing from the cystic cavity producing a foul smell.

Histopathological examination showed a lobular lesion composed of proliferating vascular network associated with fatty tissue and varying amount of myxoid stroma (Figs. 3 and 4). The latter stained with Alcian blue at pH2.5 and did not stain with PAS (Fig. 5). Proliferative index was low at <1% by Ki-67. There was no evidence of cellular atypia or malignancy. The postoperative period was uneventful and the patient was discharged on the same day of surgery. No recurrence has been seen at 6-month follow up.

3. Discussion

Lipomas are most common soft tissue tumors. Approximately 15% arise in the head and neck, especially superficial and subcutaneous layers [17]. In contrast, our case arose in much deeper layers, masquerading as a cyst rather than a solid mass.

Lipomas may be hereditary or can arise de novo. The latter are seen in obesity, diabetes, radiation exposure, endocrine disorders, insulin injection, corticosteroid therapy and trauma. There was no family history in our case and no predisposing factor as in aforementioned, was elicited.

Although lipoma is a common benign tumor, AML subtype is rare. First description of a case [2] was in a man with testicular swelling that later proved to be spermatic cord tumor. The tumor consisted of proliferating mature adipocytes associated with a myxoid stroma and numerous blood vessels [2–15]. After this first case,
19 others were reported in different sites [2–16]. The most frequent location was in the extremities. Patients were aged 4–69 years. Our 78-year old patient exceeded this range.

Use of ultrasonography, computed tomography (CT), magnetic resonance imaging (MRI), and fine-needle aspiration (FNA) have been reported to aid in planning surgical management. Radiological features of AML can mimic lipoma, heterogeneous adipose tissue and can masquerade as nonspecific or cystic mass [1]. MRI is the best diagnostic radiological method that can accurately identify lipoma and its variants preoperatively by comparing signal intensity on T1- and T2-weighted images. In fact, MRI in our case showed typical signal intensity patterns with high signal intensity on T1- and T2-weighted images without contrast media enhancement and with a weak signal on fat-suppressed images. A feature that cannot be elicited by a CT scan.

Gross appearance as a cyst in our case was interesting; its softness led to rupture, defying gentle handling during surgery. Whenever faced with another case, the surgeon should be forewarned of its unusual friability.

Histology confirmed the diagnosis in our case. Lipomas contain fat cells, while subtypes such as angiolipoma, angiomylipoma, leiomyolipoma, osteolipoma and chondrolipoma contain other types of mesenchymal tissue [12,13,15].

Histological findings in our case were classical. Cytochemical stains for the myxoid substance were thought sufficient for identifying tumor components, since lipomatous and vascular elements were self evident. Proliferative index by Ki-67 had been tested in two previous cases. In both cases, Ki-67 was under 3% [7]. In our case, it was below 1%.

Differential diagnoses include benign and malignant lesions. AML should be differentiated from myxoid liposarcoma which is highly malignant and much more common than AML with different prognostic and therapeutic implications. In some cases, myxoid liposarcomas are well surrounded by fibrous tissue with scanty mitoses and mild cellular atypia. In such cases, presence of lipoblasts and a plexiform or “chicken wire” vascular pattern distinguish them from AML. If a tumor is hypervascular, the alternative possibility of low-grade myxofibrosarcoma should be considered. However, AML lacks cell pleomorphism, nuclear atypia, hypercellularity with no curvilinear vascular pattern seen in low-grade myxofibrosarcoma. Benign lesion such as spindle cell lipoma may be considered in the differential diagnosis.
4. Conclusion

Angiomyxolipoma is a rare variant of lipoma. Cases were most frequently reported in extremities. Precise diagnosis is important to avoid unnecessary investigations, stress and misdiagnosis of myxoid liposarcoma. Our case, being the oldest (78 year-old) ever reported, is the first to have facial location and myxoid content reacting to connective tissue mucin stain. Awareness of the diagnosis should be stressed.

Conflicts of interest

None.

Funding

None.

Ethical approval

Consent form, which was signed by the patient.

Consent

Informed consent was signed by the patient.

Author contribution

Sami AL-Bdairat: study concept, data analysis, Isam Alrawi: Data analysis and interpretation, Mohammad Obaid: Manuscript preparation and editing, Yahia dajani: data collection, quality control.

Registration of research studies

This is case report not a clinical study.

Guarantor

Sami AL-Bdairat.

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