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Case Report Open Access

Giant Lipoma of the Shoulder: A Conundrum in a Child

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Abstract

Lipomas are the most common soft tissue tumors that are encountered in clinical practice; however, the giant lipomas are extremely uncommon and are entities of great concern as they can often create a diagnostic dilemma and therapeutic challenge owing to their size, location and their close relationship with other benign and malignant mesenchymal neoplasms. These tumors are rarer in children as compared to the adults, with very few cases reported in the surgical literature worldwide. We herein present a case of a giant lipoma of the right shoulder in an 11 y old boy which was diagnosed on cytology as well as on radiology and successfully managed by complete excision.

Keywords: Children, Giant lipoma, Liposarcoma, Shoulder.

Introduction

Lipoma is the most common benign soft tissue mesenchymal neoplasm, arising from the mature white adipocytes. Epidemiologically, it is frequently found in obese individuals especially females between 40 to 60 y of the age [1] and rarely it occurs in children. Nevertheless, adipose tissue tumors constitute 6% of soft tissue tumors in paediatric population [2]. Its exact etiopathogenesis is still debatable, however hereditary conditions, such as familial multiple lipomatosis and Bannayan-Riley-Ruvalcaba syndrome (BRRS), adiposis dolorosa, Gardner's syndrome and Madelung's disease may include lipoma as one of its components [3-6]. Genetic studies have revealed that HMGIC (*HMGA2*) gene, encoding a family member of the high mobility group of proteins, located at 12q15 is affected in some lipomas [7]. Few researchers have also documented a controversial link between trauma and the development of lipomas [8,9]. Lipoma has a tendency to occur at almost any anatomical location of the body where fat normally exists, therefore it is also known as a ubiquitous or universal tumor [10]. A conventional lipoma can arise within subcutaneous tissue (superficial lipoma) or within deep soft tissues (deep lipoma) or even on the surfaces of bone (parosteal lipoma). Lipomas are usually small solitary asymptomatic lesions and rarely grow to an exceptionally symptomatic large size. Superficial lipomas are generally smaller (<5 cm) than the deep seated ones (>5 cm) and approximately 5% of patients have multiple lipomas. A lipoma is considered giant when it is greater than 10 cm in any dimension or weighs more than 1000 g [11]. We herein describe a rare case of a giant superficial lipoma of the right shoulder in a child so as to create awareness among the dealing clinicians about this enigmatic entity.

Case Report

An 11 y old boy belonging to a rural background presented to the orthopaedic outpatient department with a huge swelling of the right shoulder region. The swelling was present since the past 3 years. It was insidious in onset and was of small size when the parents of the child noticed it first; however it gradually increased to attain the present size. There was history of difficulty in sleep and restricted right shoulder movements since last one month. There was no history of pain, fever, loss of appetite or weight, trauma, difficulty in breathing and upper limb weakness. His birth, developmental, personal, medical and family histories were non-contributory. General and systemic examinations were unremarkable. On local examination, a large swelling was present over the right shoulder. It was approximately 18 × 13 × 10 cm in dimensions. The overlying skin showed prominent congested blood vessels without any discoloration. On palpation, the swelling was non-tender, firm in consistency and the surface was lobulated. There was no increase in the local temperature and it was not fixed to the overlying skin and surrounding structures (Figure 1). There was no regional lymph node enlargement. Movements of the right upper limb were within the normal limits except for some discomfort while doing the abduction of the arm. Sensory and motor examination of right upper limb was normal. His routine laboratory parameters were within the normal limits. A provisional clinical diagnosis of a soft tissue malignancy was made. Fine needle aspiration cytology (FNAC) of the swelling yielded oily aspirate, which when smeared onto a clean glass slide had a glistening appearance. Microscopic examination of the May-Grunwald-Giemsa (MGG) stained FNAC smears revealed three-dimensional adipose tissue fragments with delicate blood vessels in between them. Individual fat cells were univacuolated with small round to ovoid nuclei pushed to the periphery. No nuclear atypia, lipoblasts or skeletal muscle was seen (Figure 2). Based on these cytomorphological features, a diagnosis suggestive of a giant lipoma was rendered. On radiological workup of the child, chest X-ray revealed a soft tissue shadow around the right shoulder with normal bones. Magnetic resonance imaging (MRI) scan of the right shoulder exhibited a large, lobulated hyperintense mass in the subcutaneous plane of the right shoulder region measuring 19 × 13 × 11 cm in size with multiple hypointense septae within it. It was seen to be insinuating between the right triceps and deltoid muscle. No evidence of any necrosis or calcification was seen. The glenohumeral as well as acromioclavicular joints, rotator-cuff tendons, shaft of the humerus and neurovascular bundles of the right arm were normal (Figure 3).

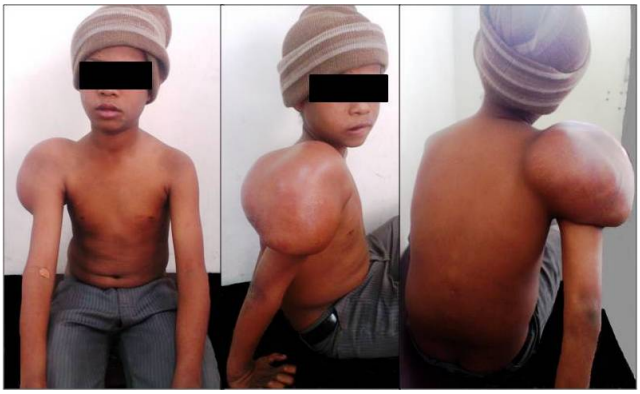

Figure 1: Giant lipoma of the right shoulder on clinical inspection.

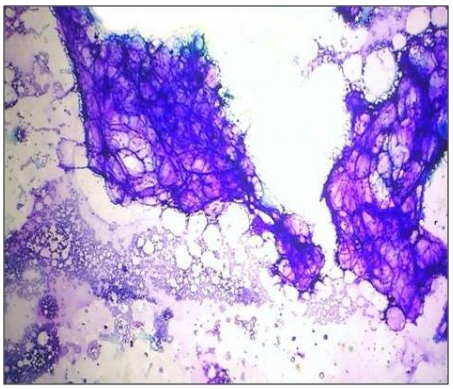

Figure 2: Mature adipose tissue fragments on cytology (MGG, X200).

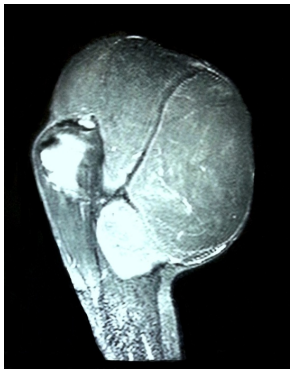

Figure 3: MRI showing a large hyperintense mass with radiating bands.

The radiological findings were consistent with the cytological diagnosis of a giant lipoma. The child underwent surgery after proper pre-anesthetic check-up. The mass was excised completely and sent for histopathological examination. Grossly, a well-circumscribed, encapsulated greyish yellow soft tissue piece measuring 19 × 13 × 11 cm and weighing 800 g was received (Figure 4a). On cut section, it was homogeneous, greyish yellow and greasy on touch (Figure 4b). Histopathology sections showed an encapsulated lesion comprising of lobules of mature adipose tissue, separated by the fibrous septa, hence confirming the radio-cytological diagnosis of a giant benign lipoma (Figure 5). The postoperative period of the child was uneventful. At the two-month follow-up, the child is doing well with normal movement of the right shoulder and there are no fresh complaints.

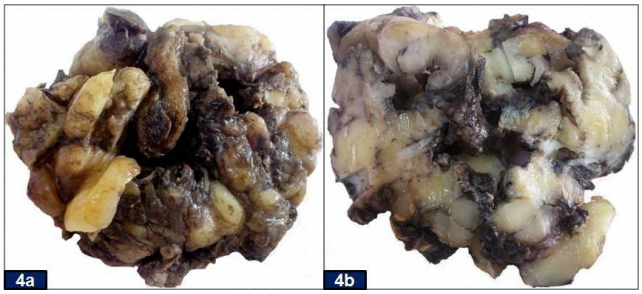

Figure 4: Resected giant lipoma (a) the encapsulated tumor (b) homogenous yellow cut surface.

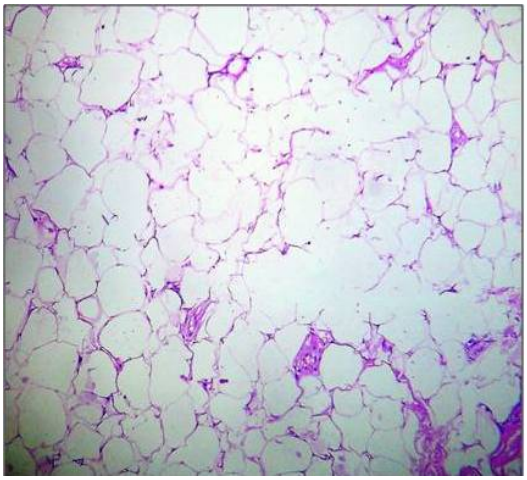

Figure 5: Histopathological sections exhibiting mature adipocytes, varying slightly in size and shape and having small eccentric nuclei (Hematoxylin and eosin, X200).

Discussion

Giant lipoma is an uncommon entity which has been seldom reported in children [2,12]. It is mainly seen on the thighs, shoulder and the trunk. The clinical features of these giant lipomas are mainly due to their size and may include pain from the stretching or compression of the adjacent nerves, compartment syndrome, restriction in the movements of the joint involved, difficulty in sleep and social embarrassment [13,14]. In the present case, the tumor was located on the right shoulder and measured 19 × 13 × 11 cm in dimensions, thus falling in the category of giant lipoma. There was a delay in the presentation of the child to the hospital owing to the negligence of the parents as they belonged to a rural setup, leading to advancement of a small lipoma to a giant one.

The main concern while dealing with giant lipomas is to rule out malignancy. Liposarcoma is the most common soft tissue malignancy in the long standing lipomas; however, such occurrence is very rare [15]. It constitutes approximately 10% of all the soft tissue sarcomas and is predominantly a disease of adulthood with peak incidence around the 5th to 6th decade of life and with a slight male predominance. The extremities are the most common primary site, accounting for about 40% of the cases. Although all subtypes are most common in the extremities, a significant proportion of pleomorphic and well-differentiated tumors arise in the retroperitoneum and in other visceral spaces. In contrast, the pediatric liposarcoma has a different spectrum of presentation compared to the adults [16]. They are extremely uncommon childhood tumors, representing about 2% of all the childhood soft tissue sarcomas, with the peak incidence occurring in the second decade of life [17]. Myxoid liposarcoma is the most common histological subtype encountered in children, usually occurring in the extremities and the overall prognosis for myxoid tumors is excellent, generally with surgical treatment alone. Pleomorphic liposarcoma occurs in axial sites, and despite multimodal therapy, the outcome is poor. Another concept documented in regard to liposarcomas is that the histopathological features of dedifferentiation are the hallmark of malignant change in a benign lipoma. Dedifferentiated liposarcomas occur most frequently in the sites in which there are chances of delayed diagnosis such as retroperitoneum. Therefore, malignant changes in giant lipomas are most commonly encountered in the retroperitoneum [14,15,18].

Histopathological examination of the resected specimen is the gold standard for a definitive diagnosis of giant lipoma. Nevertheless, once it is suspected clinically, other investigations can aid in the diagnosis by providing additional information about the tumor. The characteristics of benign lipomas on ultrasonography, computed tomography and MRI scan have been well established, and even technetium-99 diethylenetriaminepentaacetic acid scanning as well as FNAC including liquid based cytology has also been used to confirm the diagnosis [18-22]. MRI is the preferred modality for the evaluation of a soft tissue mass after X-ray films have been taken. Features that suggest malignancy include old age, large size, presence of thick septa, presence of nodular and/or globular or non-adipose mass like areas, and decreased percentage of fat composition [23,24]. In our case, both the FNAC and MRI scan confirmed the benign nature of the lipoma.

Lipomas in their typical form, rarely present a diagnostic problem for the pathologist. However, deep lipomas (intramuscular or intermuscular lipoma, perineural lipoma) or those with unusual features (chondroid lipoma, hibernoma, cellular angiolipoma, and spindle cell/pleomorphic lipoma) may be confused with liposarcomas. Therefore, on cytology and histopathology, it is important to observe appropriate background and prevent the diagnostic pitfalls as liposarcomas are associated with immature fat cells or lipoblasts. These cells have an eccentric, hyperchromatic nucleus that is indented or scalloped by the presence of one or more fat vacuoles but sometimes the lipid-laden macrophages may mimic lipoblasts on cytology and other similar cells can be seen in a variety of non-lipomatous lesions (silicone reactions). Failure to identifying such cells can lead to the overdiagnosis of liposarcoma [14,25]. Surgical excision is the treatment of choice for these giant lipomas due to their size, tendency to recur and the potential hazard of malignant transformation [26]. Blunt dissection along with an optimal hemostasis usually serves the purpose and preserves the surrounding structures as well. Dead space created following the dissection of a giant lipoma is usually drained using a suction drain to avoid the formation of hematomas or seromas. Another option for the treatment is liposuction, which is done if the malignant potential has been excluded [27,28].

Conclusion

Giant lipoma is a rare tumor which can occur in any age group and can have varied presentations, thereby creating a diagnostic dilemma. Therefore, it is essential for the dealing clinicians to be familiar with the epidemiological and anatomical attributes of the giant lipomas since a long-standing giant lipoma may resemble a malignant lesion. Nevertheless, a meticulous workup should always be done to rule out the possibility of liposarcoma in such cases. Detailed clinical examination, radiology, preoperative cytology is important before deciding on appropriate surgical management. However, it is the histopathology which is important for a definite diagnosis which is usually followed after the adequate open surgical excision of these giant lipomas. Children presenting with lipomas should also be examined carefully to look for the other signs of syndromes like BRRS, because they may need further follow-up for tumor screening in adulthood. A repeat examination over time to monitor for recurrence is mandatory as any lipomatous mass may recur especially with an incomplete excision and can undergo a malignant transformation.

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