Adult-Onset Angioblastoma of Nagakawa: A Rare Case Report

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Abstract: Tufted angioma is a rare vascular tumor commonly localized in the skin and subcutaneous tissue and is characterized by slow angiomatous proliferation. It was first recognized in Japanese literature as “Angioblastoma of Nagakawa”, a benign progressive angioma, with a variable clinical presentation. Herein, we report a case of an adult-onset Angioblastoma of Nagakawa or Tufted angioma localized over the right upper abdomen in a 41 year-old male. This case report demonstrates a rare incidence. As evident by this case report, tufted angioma although not described over the abdomen before, should be considered as a differential diagnosis of other benign vascular lesions.

Keywords: Tufted angioma, Angioblastoma of Nagakawa, Progressive capillary hemangioma

1. Introduction

Tufted angioma is a rare vascular tumor commonly localized in the skin and subcutaneous tissue and is characterized by slow angiomatous proliferation. It was first recognized in Japanese literature as “Angioblastoma of Nagakawa”, a benign progressive angioma, with a variable clinical presentation. Angioblastoma was first reported by Nagakawa in 1949. Wilson-Jones reported a case under the name “Tufted angioma (TA)” in 1976 that seemed to be almost the same disease as angioblastoma. Since then, many similar cases have been reported as tufted angioma in Europe and the United States. Wilson-Jones and Orkin coined the term “Tufted angioma” for this unusual pattern of angiomatous proliferation which was found to have some morphological similarities with strawberry nevi. Many authors have concluded that acquired tufted angioma described by Wilson-Jones and Angioblastoma of Nagakawa are the same. However, as the name suggests, the lesion is described as cannon ball like, small circumscribed angiomatous tufts and nodules in the dermis and subcutaneous tissue with characteristic lymphangioma-like vessels and pattern. Tufted angioma can be congenital or acquired, commonly presents in infancy or early childhood, can be present at the time of birth in approximately 25% of cases. A few cases of Tufted angioma have also been reported in adults. They typically occur early in life, usually involve the trunk or neck, and progressively enlarge. Although the lesions may involve a large area in some patients, they are benign, and malignant change does not occur. Tufted angiomas commonly present as a macule, papule or plaque over the upper trunk, neck and proximal part of the limbs, however involvement of face, oral mucosa and lip are also known.

Histologically, both congenital and acquired (adult onset) tufted angiommas demonstrate vascular tufts of tightly packed capillaries, randomly dispersed throughout the dermis in a typical “cannon ball” distribution with numerous crescent spaces surrounding the vascular tufts and lymphatic like spaces within the stroma of the tumor. Tufted angioma may persist unchanged or regress completely within a few years. Involvement of abdomen with Tufted angioma is previously underreported. Herein, we report a case of an Adult-onset Angioblastoma of Nagakawa or Tufted angioma localized over the right upper abdomen in a 41 year-old male.

2. Case Report

A 41 year-old male presented to the OPD with a history of an insidious onset, gradually progressive, painless swelling over the right upper abdomen (right hypochondrium) of 5 years duration. History of presence of similar lesion over the left thigh for which he had undergone surgical excision for the same 20 years back. Details of the lesion and surgical procedure were not available with the patient. There was no history of trauma, abdominal pain. No history of bowel and bladder disturbances. There was no family history of similar lesions. Upon arrival to the OPD, his vital signs were as follows: temperature 97.6 degrees Fahrenheit, heart rate 76 beats per minute, saturation 100% on room air and Blood pressure 124/78 mm Hg. He was conscious and oriented. On systemic examination: CVS: S1S2 heard, RS- Air entry bilaterally equal with no added sounds. Rest per abdomen examination was normal. Blood investigations were under normal limits. Human immunodeficiency virus (HIV) serology, Hepatitis-B and...
Hepatitis-C viral markers were negative. Chest X-ray and ECG were normal. Examination revealed a purplish, hemispherically elevated, compressible, elastic, non-tender, fixed on the superficial and deep tissues, nodular lesion measuring 5x4cm having a smooth surface localized over the right hypochondrium. A wide local excision of the lesion was performed under general anaesthesia. At the excision, the lesion appeared to be reddish-purple and ill-defined with intermingling soft and firm areas. It was then submitted for histological examination. Histological examination revealed skin tissue comprising of epidermis and dermis with epidermis showing mild focal hyperkeratinisation with different areas of atrophic changes and loss of rete ridges. The lower dermis showed a fairly circumscribed lesion lined at places by thin fibrous tissue which was composed of closely packed vascular spaces arranged in lobular pattern with varied morphology including many of them forming linear, elongated branching structures reminiscent of staghorn like vascular spaces. The vascular spaces were lined by flattened attenuated endothelial cell with occasional focal areas showing mild increased cellularity. Few rounded structures with tufting of capillary like structure reminiscent of glomeruloid bodies were also noted. Occasionally thick walled degenerated blood vessels surrounded by compactly arranged spaces forming vascular structures of varying shapes lined by attenuated oval to spindle cells also noted. A diagnosis of “Acquired (Adult-onset) Tufted angioma or Angioblastoma of Nagakawa” was made based on the above histopathological findings. Thus a large surgical excision was performed with a satisfactory cosmetic result. The patient was discharged on the same day and there has been no recurrence after more than 1 year of follow-up.

3. Discussion

Tufted angioma, also called “Angioblastoma of Nagakawa” is a rare, slowly progressive benign vascular tumor which can have variable clinical morphology. The name “Tufted angioma” was first proposed by Wilson-Jones in 19766. A similar lesion had been previously described by Milan and Champion in 1971 as a progressive capillary hemangioma5. Originally, tufted angioma was introduced as a class of disease to be differentiated from Kaposi’s sarcoma. Wilson-Jones chose the name “Tufted angioma” to avoid confusing “Angioblastoma” with the hemangioblastoma that occurs in the cerebellum. Until now only 157 cases have been reported in the English literature5. Kumakiri et al. observed two cases of angioblastoma under electron microscopy and found characteristic findings of dense bodies approximately 1 micron in size in the cytoplasm of endothelial cells7. High power observation showed the presence of crystalline lamellae consisting of alternating double and broken lines at 20 nanometre intervals. Not all reporters, however, have found these crystalline lamellae. Tufted angioma can present as a red or bluish to violaceous papule, plaque or nodule over neck, upper back, trunk and proximal part of limbs typically in childhood. Sometimes they are seen as papules, or papules developing within macular lesions. They usually range in size from 2cm to 5cm but may be much larger and induration is frequently noted within lesions. The lesion grows slowly and insidiously, and usually stops growing after some years with little tendency of spontaneous regression. These lesions may also develop in adults or the elder population. There is no sex predilection. Most series claim that 60-70% of Tufted angiomas develop before the age of 5years, and 25% of the tumors appear before the age of 50years, and its onset in people older than 60-80years is relatively rare. Cases seem to appear sporadically have been reported. Tufted angioma is generally described as a dull red, brownish-red, or purple, cutaneous patch, plaque or a nodule with angiomatous appearance. It slowly enlarges for 5 months to 10 years, after which no further growth occurs. The lesions range from few millimetres to few centimetres in size but may be extensive covering larger areas and can be even multifocal. On palpation, tufted angioma often has a rubbery consistency and may be tender. Some lesions may resemble connective tissue abnormalities. Most of the lesions are asymptomatic but may present with tenderness, hypertrichosis and hyperhidrosis. Tenderness, hypertrichosis and induration can be useful in differentiating Tufted angioma from common hemangioma. Microscopically Tufted angioma has a classical morphology. It is found to disperse in the dermis and extends into the subcutaneous tissue and is seen as discrete round to ovoid angiomatous aggregates composed of relatively bloodless, poorly canalized capillaries. Capillaries are lined by plump endothelial cells, which may show slight spindling. The endothelial cells show reactivity for several markers, including CD31, CD34 and Von-Willebrand factor (Cloting factor VIII). Pericytes are seen surrounding the capillaries and are the predominant cellular component of Tufted angioma. These cells have indistinct cell boundaries, scant cytoplasm, and oval to slightly elongated nuclei with bland morphology. Dilated crescent shaped lymphatic like vascular channels are seen surrounding the angiomatous lobules lined by plump to flattened cells with oval to slightly spindly nuclei. Clinical differential diagnosis includes eccrine angiomatosus hamartoma, port-wine stain, morphea, lipogranuloma, connective tissue nevus, appendageal tumor, sarcomoidosis, foreign body granuloma, necrobiosis lipoidica, diabeticorum, Kaposis’s sarcoma, Kimura’s disease, sarcoma, Kaposiform hemangioendothelioma and especially adult-onset hemangiomas. The pathogenesis of Tufted angioma is unknown. No causes of Tufted angioma have yet been established. Trauma does not appear to be a predisposing factor, although some report describes the development of a lesion of Tufted angioma at the site of a previous arthropod bite. Some authors have noted the development of Tufted angioma within port-wine stains, others have postulated that high hormonal levels during pregnancy and puberty may induce the development of Tufted angiomas8. Perhaps localized secretion of growth factors prompts development of numerous tufts of vessels. Growth factors such as Interleukin-8 affect angiogenesis. Tufted angioma may be associated with the platelet-trapping syndrome or Kasabach-Merritt syndrome; the presence of petechial and ecchymotic patches should alert the physician to the development of Kasabach-Merritt syndrome. No specific laboratory study is useful in the diagnosis or treatment of Tufted angioma. If the
coexistence of Kasabach-Merritt syndrome is suspected, a complete blood cell count with a determination of platelet count, prothrombin time and activated partial thromboplastin time and full disseminated intravascular coagulation profile is indicated which may be deranged. Many treatments have been used for Tufted angiomas, surgical excision was successful in some lesions. Dye laser treatment protocol produced a satisfactory result in some patients. However, recurrences after surgery are common. Cryosurgery, X-ray therapy, and electrocautery have been tried with recurrence of lesions. Treatment with topical and intrallesional corticosteroids for an actively growing lesion had no effect. Unless the lesion is symptomatic or cosmetically disfiguring, observation is the treatment of choice for lesions that are not amenable to excision. If removed completely, the lesion does not recur. Despite the progressive spread of the angiomas, they appear to be benign. Malignant transformation of a Tufted angioma has not been reported yet.

4. Conclusion

This case report demonstrates a rare incidence. As evident by this case report, tufted angioma although not described over the abdomen before, should be considered as a differential diagnosis of other benign vascular lesions. Tufted angioma can have variable clinical presentation and can present at any age. A complete physical examination and haematological work up has been recommended in patients with classic Tufted angioma to exclude rare association of Port-wine stain and Kasabach-Merritt syndrome with this rare entity.

References


Figure 1: Purplish hemispherically elevated, elastic, nodular lesion measuring 5x4cm having a smooth surface localized over the right hypochondrium.

Figure 2: Post surgical excision a purplish, hemispherically elevated, compressible, elastic, fixed on the superficial and deep tissues, nodular lesion measuring 5x4cm.
Figure 3: Cut section showing brownish haemorrhagic areas

Figure 4: Histopathological examination showing a circumscribed lesion lined at places by thin fibrous tissue which is composed of closely packed vascular spaces arranges in lobular pattern with varied morphology including many of them forming linear, elongated branching structures reminiscent of staghorn like vascular spaces, thick walled degenerated blood vessels surrounded by compactly arranged spaces forming vascular structures of varying shapes lined by attenuated oval to spindle cells also noted. (Stain, haematoxylin and eosin; original magnification, ×40)