

Xanthoma Masquerading as Bartholin's Cyst in Vulva

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Abstract: A case of 17-year-old female who presented with a cystic swelling on the left side labia majora progressively increasing in size for 3-4 months, which is painful for a week. Samples from the painful cystic swelling was excised, under spinal anaesthesia after obtaining relevant consent. The excised cyst was approximately 3x2 cms, sent for histopathological examination. Histopathologic examination showed collection of xanthoma cells and reported as xanthoma of the external genitalia. Lipid profile was within normal range.

Keywords: Xanthoma, vulva, Infected Bartholin's cyst, Cyst in labia, Normolipemic xanthoma.

1. Introduction

Xanthoma is a localised collection of foamy histiocytes loaded with lipid, seen in the subepithelial connective tissues of skin and tendons[1]. It is may not be a true tumor but, rather, a reactive histiocyte proliferation that occurs in response to alteration in serum lipids. Xanthomas may develop in most primary and secondary hyperlipoproteinemias and occasionally in the normolipemic state[2].

There are wide array of disorders which can display similar histologic pattern like metabolic disorders (other disorders of fat metabolism other than hyperlipoproteinemias, lysosomal storage disorders), infectious diseases due to Haemophilus organisms which gives haemorrhagic exudate, tumor (lymphomas and melanocytic nevi), trauma (insect bite, folliculitis, another insult) and idiopathic conditions.

The xanthomas may clinically be divided into eruptive xanthomas, tuberous xanthomas, tendon xanthomas, and plane xanthomas. Xanthelasmas are viewed as a morphologic variant of plane xanthoma more closely related to the macrophage disorders, but many other described it along with xanthomas for the sake of continuity. **Plane xanthomas** typically develop in skin folds and especially in the palmar creases, where they are diagnostic for dysbetalipoproteinemia. **Diffuse plane xanthomas** are typically seen as multiple grouped papules and poorly defined yellowish plaques in normolipemic patients, often with paraproteinemia, lymphoma, or leukemia. On the other hand, **intertriginous plane xanthomas** suggest homozygous familial hypercholesterolemia. The palmar xanthomas associated with cholestasis (primary biliary cirrhosis and biliary atresia) are plaque-like and tend to extend past the creases.

2. Case Report

17 year old female brought to the Obstetrics and Gynaecology OPD by her parents with the complaints of swelling in the vagina. A diagnosis of Bartholin's cyst left side was made and excision biopsy was planned. Clinically the cystic lesion measured 4x3 cms and was excised under spinal anaesthesia. Procedure done with the consent from her parent and sent to the Department of Pathology, Rajah Muthiah Medical College, Annamalai University, Chidambaram. Post operative sequelae were within normal limits. Gross examination revealed a single grey, brown grey, black soft tissue piece measuring 2.5x2x0.5 cm identified. On cut section-pultaceous material expelled out. A uniloculated cyst of uneven thickness seen with solid areas appears to be orange-yellow in color. The size discrepancy of the specimen is due to the effects of formalin fixation.

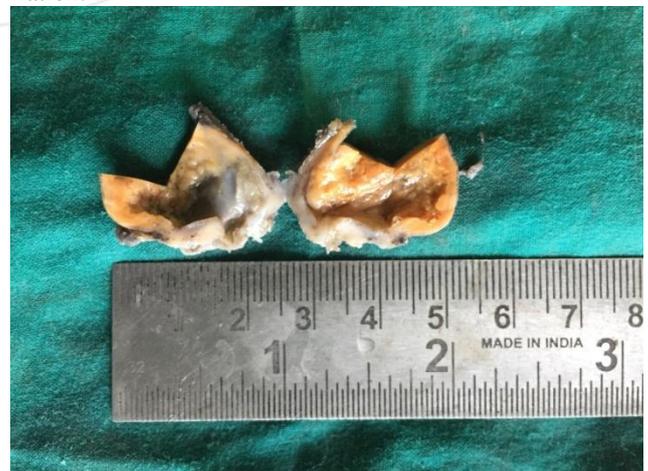


Figure 1: Gross picture showing empty cyst wall. The inner lining is orange-yellow in color. Wall is of variable thickness.

Microscopy of multiple sections studied revealed, a well circumscribed lesion, where large number of foamy histiocytes arranged in sheets were noticed. Foamy histiocytes-xanthoma cells were polygonal in nature with pinpoint nuclei and abundant cytoplasm which were foamy in nature were seen, interspersed with collection of lymphocytes and plasma cells. Extensive areas of fibrosis was also observed. Central portion exhibits liquefactive degeneration with formation of cholesterol clefts, occasionally surrounded by giant cells which were flattened. The histological features were consistent with the diagnosis of xanthoma – vulva.

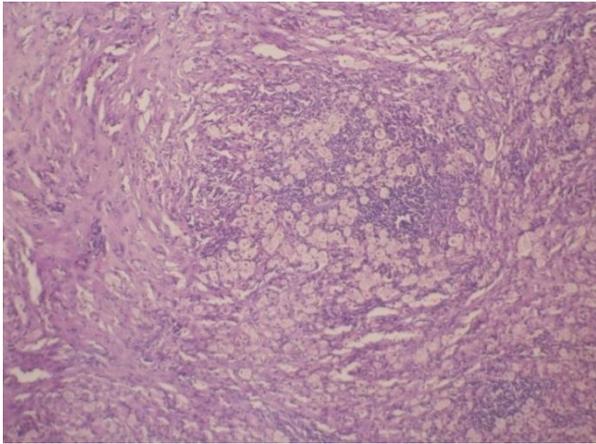


Figure 2: H&E-20x xanthoma cells along with lymphocytes and plasma cells

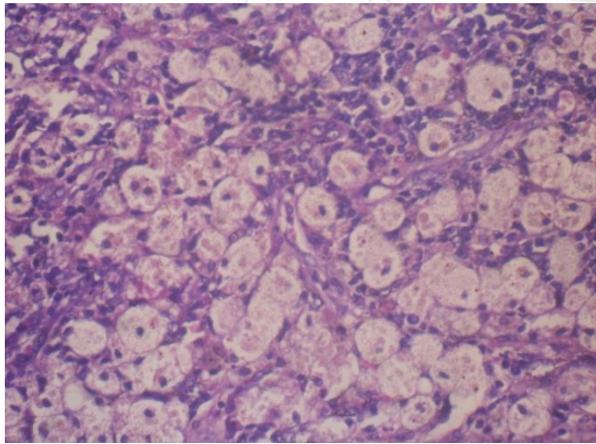


Figure 3: H&E-40x xanthoma cells with bland and small pyknotic nuclei, lymphocytes and plasma cells

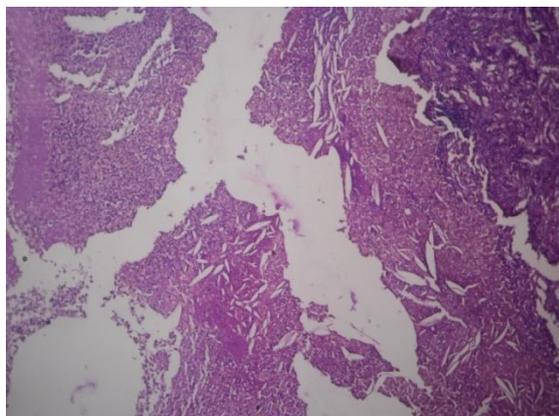


Figure 4: H&E-10x showing collection of extracellular cholesterol (cholesterol clefts)

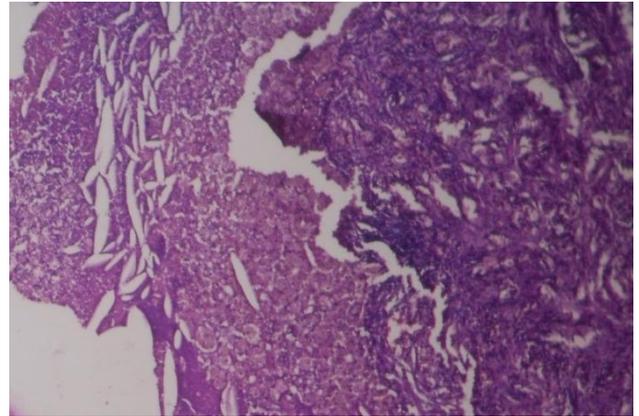


Figure 5: H&E-10x A compressed giant cell around cholesterol clefts

3. Discussion

Most patients with lipoprotein abnormalities do not have xanthomas, but the presence of xanthomas and especially the identification of extracellular lipids on histologic examination of the lesion should alert the clinician to the need for an internal evaluation. Although xanthomas can potentially occur at any soft tissue site, the localization stimulus seems directly related to the vascular permeability, as agents that increase permeability (e.g., histamine) can accelerate xanthoma formation. Minor trauma or injury that results in histamine release also accelerates xanthoma formation [3]. Histamine is one of the major vasoactive amines, which is an important mediator of acute inflammation. It is liberated in acute inflammation, and that it can mimic some of the vascular events [4]. Histamine causes dilation of arterioles and increases the permeability of venules which results in extravasation of RBCs.

Xanthomas were formerly considered neoplastic, their association with hyperlipidemic states reveals that they are reactive lesions. The lipid in xanthomas are derived from blood. This has been demonstrated experimentally that serum lipoproteins leave the vascular compartment, traverse small vessels, and enter the macrophages of soft tissue. Once ingested by macrophages the lipoprotein is degraded to lipid, and the lipid is released to the extracellular space [5]. The main source of cholesterol crystals are from the rupture of red blood cells with breakdown of the lipid layer of erythrocyte cell membrane [6].

The histologic appearance of xanthomas of the skin and the tendons is characterized by foamy cells, macrophages that have engulfed lipid droplets [7]. Most of the xanthoma cells are mononuclear, but giant cells, especially of the Touton type with a wreath of nuclei, may be found. The most diagnostic finding suggesting that a xanthomatous lesion reflects an underlying lipid abnormality is the presence of free lipids that have not yet been taken up by macrophages. The lipid droplets can be better seen if fresh frozen or formalin-fixed frozen sections are stained with fat stains such as scarlet red or Sudan red. There may be varying degrees of fibrosis, giant cells, and clefts, depending on the type and site of xanthoma sampled, but most are surprisingly similar. All xanthomas are characterized by a degree of fixation artifact. Formalin fixation and paraffin embedding

remove lipids so that only their shadows are left behind. Larger extracellular deposits of cholesterol and other sterols leave behind clefts.

4. Conclusion

Xanthomas are wide range, they are following haemorrhagic exudates, trauma in extensive areas, hyperlipidemia, rarely in normolipemic patients. There was no underlying lymphoproliferative disease and clinicopathologically there was no suggestion of xanthoma disseminatum, juvenile xanthogranuloma or generalised eruptive xanthomas. In the present patient, xanthoma is probably secondary to an ancient Bartholin's cyst. Hence, the case is documented.

References

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