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INTERNATIONAL JOURNAL OF ADVANCED RESEARCH (IJAR)

Article DOI: 10.21474/IJAR01/14642
DOI URL: http://dx.doi.org/10.21474/IJAR01/14642

RESEARCH ARTICLE

A CASE OF VUVLVAR ANGIOMYXOMA : REPORT AND LITERATUREREVIEW

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Manuscript Info

Manuscript History
Received: 28 February 2022
Final Accepted: 30 March 2022
Published: April 2022

Abstract

Superficialangiomyxoma (SMA) is a rare benign skin tumorwith a peak incidence in middle age . The diseaseiscaused by mutations in the PRKAR1A gene. Which, codes for the type of I alpha regulatorysubunit of the enzyme protein kinase A. We report a rare case of vulvarangiomyxomain 19-year-old patient beneficied of large exicion of mass in the leftlabia majora. the diagnostic of SMA retained, afterhistologicalstudy of surgicalspecimen , The evolution has been good withoutrecurrence .

Introduction:-

Superficialangiomyxoma (SMA) is a rare benign skin tumor with a predilection for the head, neck and trunk. SMA is more common in men, with a peak incidence in middle age [1]. It was initially described as a component of the Carney complex (CC), a rare disease characterized by the triad: pigmented lesions of the skin and mucous membranes, cutaneous and cardiac myxomas and multiple endocrine tumors [2]. Now, sporadic AMS is also recognized. Due to its benign, cyst-like appearance, vulvar SMA is often confused with skin tags, labial cysts, or Bartholin's or Gartner's duct cysts[1]

Case Report :

A 19-year-old patient with no particular pathological history presented with a pedunculated mass in the left labia majora of 1 x 1 cm without inflammatory signs or ulceration opposite (Figure 1).

Figure 1: Pedunculated mass at the level of the large left lip of 1 x 1 cm without inflammatory signs or ulceration opposite.

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The conduct was excision of the lesion. Histologicalexaminationfound a superficial (cutaneous) angiomyxomawithcompleteresection (figure 2).

Figure 2:- Overview of the vulva after complete excision.

The evolution has been good withoutrecurrence to date.

Discussion:-

This case is of a patient described with vulvar AMS, a condition more frequently diagnosed in middle-aged male patients and located in the head or trunk region [1]. Vulvar SMA is rare, with a total of around 20 cases reported worldwide [3].

Patients diagnosed with AMS should be carefully screened for other manifestations of Carney Complex (CC) so that they can be fully evaluated for the various conditions associated with the disease. CC is a rare disease that is inherited either in an autosomal dominant manner or, less commonly, sporadically as a de novo genetic mutation. The disease is caused by mutations in the PRKAR1A gene which codes for the type I alpha regulatory subunit of the enzyme protein kinase A [4]. A diagnosis of CC is more likely in patients who present with multiple cutaneous myxomas in common locations such as the eyelid, external auditory canal, nipples and genitals, in association with other signs of the syndrome, such as pigmented skin lesions and endocrinopathies [4] In particular, CC patients should be evaluated for myxoma, a diagnosis that can be life-threatening if not identified in time. Cardiac myxomas are the most common non-cutaneous lesions in patients with CC; they are present in 20 to 40% of patients diagnosed with the disease [4]. Early detection of cardiac myxoma is essential to avoid potentially catastrophic complications, including intermittent obstruction of blood flow, thromboembolic events, and complete occlusion of valvular flow in the heart that can lead to sudden death. a diagnosis that can be life-threatening if not identified in time.

Various endocrinopathies are possible in patients with CC, ranging from elevated levels of growth hormone, growth factor insulin, and prolactin due to pituitary tumors. Patients may also have elevated cortisol levels secondary to adrenal disease. CC is also associated with the development of thyroid nodules and thyroid carcinomas, as well as testicular and ovarian tumors. The most commonly reported ovarian neoplasms in CC patients are cystadenomas and mature cystic teratomas. Rarely, ovarian carcinoma has been reported [4]. Due to the wide range of potentialphenotypesaffected in patients with CC, comprehensive screening isrecommendedannually.

Clinically, AMS ranges from polypoid to papulonodular. Distinctive histological features of SMA include a poorly circumscribed tumor with a multilobular growth pattern. Component cells include sparse, bland, spindle-shaped cells suspended within a myxoid stroma [7]. When present, neutrophils in the cellular medium are an important diagnostic feature that distinguishes SMA from other myxoid tumors. The blood vessels inside the tumor are usually small and thin-walled. Nuclear atypia and hyperchromasia are rare [3]. Recurrence rates of AMS are reported to be 30 to 40%. It isthoughtthatrecurrences are probably due to incompleteresections [8,9] .Fortunately,It is essential to distinguish SMA from aggressive angiomyxoma (AMA). AMA is a rare soft tissue tumor with a predilection for the pelvic and perineal regions. Therefore, as with SMA, the lesions are often mistaken for Bartholin's gland cysts,

lipomas, hernias, or abscesses due to their location and extremely benign appearance. Symptoms of AMA are usually nonspecific and related to the mass effect of the lesion on surrounding structures. Local invasion of AMA into surrounding structures such as the bladder and rectum is rare; however, patients may experience morbidity due to the resection of large masses due to the need for widemargins [7].

Roughly, AMA and AMS seem to be similar; however, they show histological differences. Compared to SMAs, AMAsoften tend to beunencapsulated or onlypartiallyencapsulatedwithinfiltrativeborders and are characterized by thickenedvesselwalls [1].

Immunohistochemical staining for AMA demonstrates characteristic estrogen and progesterone receptor positivity. Additionally, they are usually positive for CD34, desmin, and vimentin. S-100 staining is usually negative [6]. Reported recurrence rates for AMA are similar to those for AMS; however, they are more likely to be locally aggressive, and metastases have been reported. Like SMA, AMA usuallyrecurs due toinadequateresectionmargins [3]. Management of AMA homes aroundsurgicalresection.

Experimentalchemotherapy has been used in selectedrecurrent cases ; however, this practice is not wellestablished. The use of hormonal modulatingdrugs for the treatment of AMA has also been attempted in some patients due to the presence of positive estrogen and progesteronereceptorswithsomesuccess.

Conclusion:-

The presentation of our patient with a large vulvar mass compatible with SMA is remarkable due to her young age and the location of the lesion. MSA is an important pathological diagnosis to identify due to the presence of associated conditions that can have significant and life-threatening impacts on a patient'shealth.

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