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Rare Case Report and Review of Literature of Tumoral Calcinosis

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Abstract: ***A 28 year old female patient was presented to the surgical opd with complaint of several days of severe and constant pain over left gluteal region. There were no relieving or instigating factors. The pain was so sharp. The patient denied any history of trauma to site. On physical examination, the patient was moderately dehydrated with normal pulse (76 pulse / min) and had normal blood pressure. The local examination showed signs of infllamation, hyper pigmentation and hardness and local tenderness. Laboratory studies were unremarkable. MRI study shows irregular area (3 x7.6 x 14cm) of heterogenously hyperintense on T1 weighted images which appears heterogenously hypointense on STIR images noted in subcutaneous plane at left gluteal region-suggest FAT NECROSIS. PD/STIR hyperintensity noted involving underlying left gluteal maximus muscle- suggest oedema. Debridement and Excisional Biopsy performed and sent for analysis. Report suggests histological features favour calcinosis. And adviced for PTH, S.Calcium, Phosphorus, S. Creatinine. All investigations were unsignificant. For Definative management lesion excision and limberg flap plasty operation performed. Total operative time was 120 minutes. Excised lesion sent for histopathological examination. The patient's subsequent hospital course was uneventful. She was discharged in satisfactory condition 10 days postoperatively. Biopsy report suggests histological features of acute on chronic inflammation, Fat necrosis, infarction, marked calcification and dystrophic bone formation in dermis, subcutaneous, fibrous and muscle tissue. No evidence of parasite and granuloma.***

Keywords: Tumoral calcinosis, phosphate, blood

1. Case Introduction

Tumoral calcinosis is an inherited condition that causes high levels of phosphate in blood. In tumoral calcinosis the minerals calcium and phosphorus build up in tissues. Body needs phosphorous and calcium to build strong bones and teeth. But when you have an excess of these minerals, they form non cancerous (benign) lumps in tissue that resemble tumors. Other term is hyperphosphatemic familial tumoralcalcinosis (HFTC).

2. Report

- A 28 year old female patient was presented to the surgical opd with complaint of several days of severe and constant pain over left gluteal region. There were no relieving or instigating factors. The pain was so sharp. The patient denied any history of trauma to site. On physical examination, the patient was moderately dehydrated with normal pulse (76 pulse / min) and had normal blood pressure. The local examination showed signs of infllamation, hyper pigmentation and hardness andlocal tenderness. Laboratory studies were unremarkable.
- MRI study showsirregular area (3 x7.6 x 14cm) of heterogenously hyperintense on T1 weighted images which appear heterogenously hypointense on STIR images noted in subcutaneous plane at left gluteal region-suggest FAT NECROSIS. PD/STIR hyperintensity noted involving underlying left gluteal maximus muscle-suggest oedema.
- Debridement and Excisional Biopsy performed and sent for analysis. Report suggests histological features favour calcinosis. And adviced for PTH, S. Calcium, Phosphorus, S. Creatinine. All investigations were unsignificant.
- For Definative management lesion excision and limberg flap plasty operation performed. Total operative time was 120 minutes. Excised lesion sent for histopathological examination. The patient's subsequent hospital course was uneventful. She was discharged in satisfactory condition 10 days postoperatively.
- Biopsy report suggests histological features of acute on chronic inflammation, Fat necrosis, infarction, marked calcification and dystrophic bone formation in dermis, subcutaneous, fibrous and muscle tissue. No evidence of parasite and granuloma.

3. Discussion

- Tumoral calcinosis is an inherited metabolic disorder of autosomal recessive type. An estimated 1 in 3 people with this condition inherit a gene change (mutation) to their FGF23 gene. This gene helps cells in your bones make a protein called fibroblast growth factor (FGF) 23. Some people inherit changes to the GALNT3 and KL genes, which regulate the production of FGF23. People with normophosphatemic tumoral calcinosis inherit a mutated SAMD9 gene. FGF23 protein mutation causes kidneys to absorb too much phosphate.
- Tumoral calcinosis causes lumps of different sizes to form near one or more joints. These lumps typically appears during early childhood or early adulthood. But infants and older adults can get them, too. And having symptoms of difficulty moving affected joint, firm lumps that don’t cause pain, musculoskeletal pain in bones or joints. Lumps from tumoral calcinosis typically form just underneath your skin near joints. Most commonly affected sites are hips, elbows, shoulders, feet, wrists, jaw.
- There are two types of tumoral calcinosis. (1) Hyperphosphatemia-hyperostosis syndrome (HHS),

which causes excess bone growth and noncancerous bone lesions. (2) Normophosphatemic tumoral calcinois, which often affects people who have kidney failure. It can lead to hyperparathyroidism. .

- Treatment consists of Non surgical and Surgical management. In Non-surgical management Phosphate binding medications to lower amount of phosphate inblood. Combining phosphate binders with a medication called acetazolamide can block your body's ability to absorb phosphorus. Definative management for tumoral calcinosis is surgical management. There is no risk of the lumps becoming cancerous (malignant).
- In our case in diagnostic debridement and excisional biopsy performed and after report for definitive management of tumoral calcinosis whole lesion excision with limberg's flap plasty. Lesion sent for Histopathological examination. Total operative time was 120 minutes and our patient's postoperative period was uneventful. He was discharged on the tenth postoperative day.

4. Conclusion

In this case of a 28-year-oldfemale patient with Left gluteal tumoral calcinosis with specific symptomatology was presented. Due to the intensification of pain and difficulty in mobilization we approached a detailed diagnostic treatment of the patient and arrived at the above diagnosis. We performed open surgery, which confirmed the formation of Left gluteal tumoral calcinosis. We resolved it in the usual way, by excising the lesion and defect repaired with limberg's flap plasty. Excisional biopsy helps us make an accurate diagnosis and perform timely surgery, which was the case with our patient.

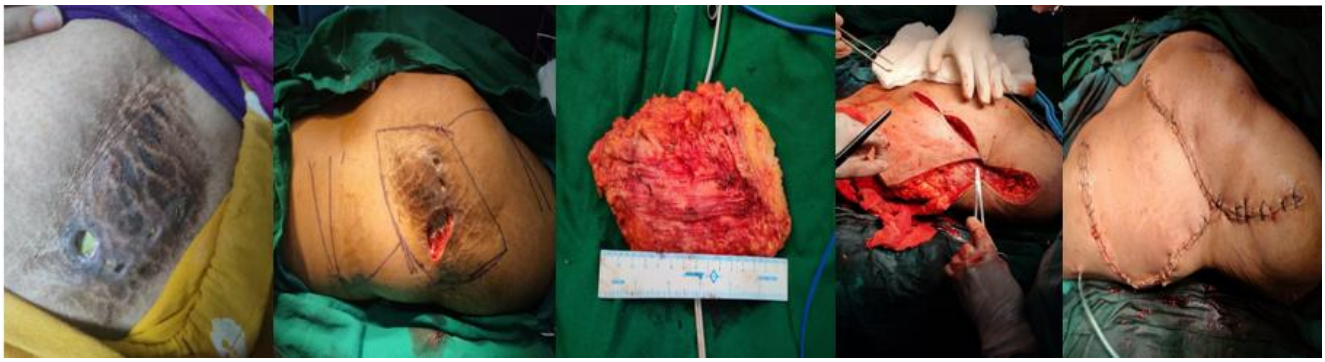

Figure 1: Lesion
Figure 2: Marking for excision
Figure 3: Excised lesion
Figure 4: Closing defect with limberg's flap
Figure 5: Post op

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