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Case Report

A rare case of perianal granular cell tumour in a 75-year-old female

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ABSTRACT

Granular cell tumours (GCTs) are a rare type of soft tissue tumour that can be found in many anatomical locations however its occurrence in the perianal region is exceedingly rare. GCT typically presents as a firm painless mass, and therefore, it may be mistaken for other more common anorectal pathology. Most are benign, however 1-2% of GCTs are malignant. Complete surgical excision is recommended for accurate diagnosis and adequate treatment. This case reports a 75-year-old female who was found to have a perianal GCT.

Keywords: Granular cell tumour, Perianal, Surgical excision

INTRODUCTION

Granular cell tumours (GCTs) are a rare type of soft tissue tumour that can be found in various anatomical locations.[1-3] Its occurrence in the perianal region is particularly infrequent with less than 30 cases reported in the literature since it was first described by a pathologist, Abrikossoff, in 1926.[2,3] GCT typically presents as a firm, painless mass. [1,3-5] As a result, when located perianally, it can lead to clinical uncertainty and potential incorrect identification as haemorrhoids, skin tags or abscess. Surgical excision is required for accurate diagnosis and adequate treatment of GCTs. Though the vast majority are benign, 1-2% of GCTs are malignant despite having very similar histopathological appearance to their benign counterparts.[2,3,6] This can be challenging for clinicians, as often the only reliable method to distinguish between benign and malignant GCT is the presence of metastases.

CASE REPORT

A 75-year-old female patient of Greek descent was referred to the colorectal surgery outpatient clinic at a metropolitan hospital in Queensland, Australia, for assessment of a perianal mass. The patient had observed the slow-growing mass for up to 12 months, and her only symptom was pruritus. She denied any changes in bowel habits or rectal bleeding. Patient was taking rivaroxaban for atrial fibrillation and had a permanent pacemaker in situ. She had no other significant medical history and was independent at home with activities of daily living.

Clinical examination of perianal region revealed a 2 cm firm, non-tender mass at 2 o'clock. Differentially diagnosis preoperatively included perianal sebaceous cyst. Patient underwent surgical excision of the perianal mass. Intraoperative findings confirmed an approximately 2 cm firm mass, which adherent to skin with superficial involvement of internal anal sphincter muscle. There was no involvement within anal canal. Mass excised in one piece and sent for histopathological examination.

Macroscopic examination found a 16×12×10 mm ovoid tan lesion, which appeared solid white on cross section. Histopathology revealed large polygonal cells with granular eosinophilic cytoplasm. Rare mitoses were identified (up to ½ mm^2) and no marked nuclear atypia or tumour necrosis was observed. Immunohistochemistry revealed positive staining with S-100. All pathology results were consistent with a GCT.

The patient has been seen in the colorectal outpatient clinic six months following surgery, and no signs of reoccurrence were observed on clinical examination.

DISCUSSION

GCTs are a rare neoplasm that can occur in a multitude of locations**,** most commonly in the head and neck, with the tongue accounting for one third of cases.[3] GCTs are also seen in the breast, bladder, respiratory tract and gastrointestinal tract (GIT).[1,3] To the best of our knowledge, there is less than 30 documented cases of GCT in the perianal region.[2,3] In 1981, Johnston and Helwig, reported 16 cases of perianal GCT, while other sources only mention isolated cases.[7]

GCT is typically a solid, non-tender polypoid mass without ulceration, usually a yellowish tan appearance.[1,6] They are often asymptomatic and found incidentally on endoscopic procedures those in the GIT. Perianal GCT are also largely asymptomatic with some cases reporting perianal discomfort.[4,5] They are slow growing and are often small lesions between 5 to 30 mm.[2] GCTs are most seen between the 4th and 6th decades of life, making our case, to some extent, atypical.[1,2] Given its rarity in this location, there is a risk that clinicians mistake perianal GCTs for benign differentials including sebaceous cyst, abscess or haemorrhoid. The only means of accurate diagnosis is histopathological and immunohistochemical examination.

Non-encapsulated, large polyhedral cells with copious eosinophilic granular cytoplasm is characteristic of GCT.[1,2,3,6,7] The cells are found in nests separated by collagen fibres and have nuclei which are usually small, centrally placed and uniform.[1] The granular cytoplasm stains periodic acid-Schiff (PAS) positive.[1,3,6] Johnston and Helwig report, in a study of 74 cases of GCT, that 50% were associated with pseudoepitheliomatous hyperplasia of its overlying epithelium, a characteristic typically seen in squamous cell carcinoma (SCC).[1,4,7] Therefore, a pathologist may mistake a rare GCT for a more commonly seen SCC, particularly if only a superficial biopsy has been collected rather than surgical excision.

The histogenesis of GCT has been debated.[7] In 1926, Abrikossoff theorised that it arose from embryonic muscle cells.[3] Over 20 different ideas have been postulated, however it is widely accepted now that GCTs are derived from the Schwann cell of peripheral nerves.[4-7] S-100 is a protein characteristic of the myelinating cell of the peripheral nervous system, the Schwann cell. GCTs typically stain positivity with S-100, as is noted in our case study.[6,7]

The recommended treatment of GCT is complete surgical excision to ensure accurate diagnosis and reduce the risk of reoccurrence.[1,3] The reoccurrence rate is thought to be between 2 and 8% for benign GCT.[1,3] The vast majority of cases are benign however 1-2% of GCTs are malignant.[2,3,6] Unfortunately, histopathological characteristics can be like one another. Concerning features for malignancy are thought to include increased mitotic count, necrosis, spindled tumour cell, prominent nucleoli and high nuclear to cytoplasmic ratio.[1] Moreover, the only reliable means of differentiating a malignant GCT from a benign GCT is the presence of metastasis.[6] GCTs have been reported to metastasise to the lymph nodes, lung, liver and bones, and have a poor prognosis with a mortality between 30%-50% at 3 years.[2] Surgical excision remains the mainstay of treatment for malignant GCT as chemotherapy and radiotherapy have not been found to improve survival.[1,3]

CONCLUSION

GCTs are rare. In the perianal region, where the differential diagnosis is broad, this case highlights the importance for surgeons and pathologists to be aware of GCTs, given it may be dismissed as other benign, more common, anorectal pathology. Due to the diagnostic challenge in distinguishing benign from malignant GCTs, complete surgical excision followed by comprehensive histopathological and immunochemical examination is imperative in the identification and treatment of all GCTs.

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