Case report

Rare presentation of a testicular glomus tumour

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SUMMARY

Glomus tumours are atypical in extracutaneous locations and very rarely located in reproductive organs. We present an unusual case of an incidentally discovered glomus tumour arising from the testis of a 47-year-old man. The testicular occurrence of this tumour type is not only exceptionally rare but also serves to highlight the debate regarding the optimal management of incidentally discovered small testicular lesions.

BACKGROUND

Glomus tumours are benign mesenchymal neoplasms that arise from the arterial segment of the glomus body or the Sucquet-Hoyer canal,^{1,2} the modified vascular smooth muscles cells involved in thermoregulatory control. The majority of reported glomus tumours are cutaneous in location, situated in tissues where glomus bodies are more densely clustered in the stratum reticulare, the skin layer lying deep to the dermis.³ Their location in other extracutaneous sites has been documented, including the liver, glans penis, respiratory system and even the heart.^{4–8} These tumours are rarely formed in male reproductive tissues, with a paucity of published literature describing their occurrence in the scrotum and testicle.^{9–11}

This is the second documented case of glomus tumour arising in the testis and aims to increase the awareness of their occurrence, advocating its consideration in the diagnosis and management of testicular neoplasms. Furthermore, the case adds to the narrative of how best to triage incidentally discovered testicular lesions.¹² Multicentre analysis of testicular lesions shows that tumour markers are unhelpful in the distinction of neoplastic from benign lesions measuring <5 mm and approximately two-thirds of the lesions measuring 5–10 mm are found to be benign.¹² For these small lesions there is an ongoing debate regarding which clinical pathway to pursue. There is a concern that proceeding to radical orchidectomy might make patients 'victims of modern imaging techniques',¹³ although with a lack of clear guidance available, monitoring versus operative management remains a decision guided by both patient preference and multidisciplinary input.¹⁴

CASE PRESENTATION

A fit and well 47-year-old man presented to his general practitioner with right-sided scrotal pain. He had no constitutional symptoms, no significant medical or family history, and no significant risk

factors for testicular cancer. Examination revealed a normal scrotum on visual inspection and normal right and left testis with no tenderness, swelling or palpable masses. There was no evidence of a hydrocele or varicocele and abdominal examination was normal. In light of the ongoing scrotal pain, an ultrasound scan (USS) was requested. This demonstrated a normal right testis. An incidental hypoechoic focal lesion, measuring 4.5×4.4×3.5 mm was found in an otherwise normal left testis. A prompt referral to urology via the 2-week wait cancer pathway was made.

INVESTIGATIONS

Due to suspicion of an evolving malignancy, surveillance was undertaken with an early repeat ultrasound (US) scheduled within 4 weeks. This US confirmed the presence of the left testicular lesion, demonstrating a solid, hypoechoic, focal lesion measuring 5.6×5×4 mm (figure 1). Vascularity was noted within the lesion with vascular distortion around the lesion perimeter. The right testicle was normal and tumour markers beta human chorionic gonadotropin (hCG), alpha fetoprotein (AFP) and lactate dehydrogenase (LDH), were all within normal range. Continued close observation was undertaken with a further USS 3 weeks later which again detected the hypoechoic focal lesion in the left testis which was unchanged in size. Throughout this period the patient remained asymptomatic. Following multidisciplinary discussion, orchidectomy was advised given the persistent nature of the lesion and the suspicion of malignancy. At his subsequent clinical appointment, the patient was fully informed regarding the treatment options including ongoing monitoring, frozen section excision (FSE), partial orchidectomy or radical orchidectomy. After careful counselling, the patient decided to proceed to a radical orchidectomy.

DIFFERENTIAL DIAGNOSIS**Germ cell tumours**

Seminoma, embryonal carcinoma, yolk sac tumour, teratoma, mixed germ cell tumour.

Sex cord-stromal tumours

Leydig cell tumour, sertoli cell tumour, granulosa cell tumour, fibrothecoma.

Other

Lymphoma, leukaemia, sarcoma, leiomyoma, vascular tumours, fibroma, neurofibroma, tuberculosis, segmental ischaemia.

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Figure 1 Ultrasound scan demonstrating hypoechoic heterogeneous lesion (5.6×5×4 mm) in an otherwise normal left testis.

Painless testicular lumps raise the clinical suspicion of malignancy. Within the spectrum of testicular malignancy, germ cell tumours account for the vast majority and are further divided into seminomatous and non-seminomatous types. In this case, the US findings also raised suspicion of malignancy given the solid nature of the lesion. Though less common, testicular lumps may be the presenting symptoms in acute leukaemia or lymphoma; hence consideration should be given to these diagnoses. Other rarer causes of testicular lesions are also noted, including vascular tumours and sarcoma.

TREATMENT

Elective left inguinal radical orchidectomy and prosthesis insertion were performed. The operation proceeded with no complications and the left testis and surrounding tissue were sent for histological analysis. The patient was discharged on the same day and re-presented briefly for a single day admission 5 days postoperatively with pain. This discomfort was controlled and the patient was discharged with outpatient follow-up 6 weeks postoperatively. CT chest-abdomen-pelvis (CT CAP), performed postoperatively to complete staging, showed no evidence of metastatic disease.

OUTCOME AND FOLLOW-UP

Gross histological examination of the left testis revealed a well-circumscribed pale-yellow lesion, measuring 5 mm in maximum dimension. Following staining and magnification, the lesion was composed of round and oval shaped cells arranged in nests and cords indicative of a glomus tumour (figure 2A,B). The concentric arrangement of tumour cells around thick-walled

blood vessels was also noted and lesional cell nuclei were bland with a fine granular chromatin pattern. No crystals were seen in the cytoplasm and there was no mitotic activity or necrosis. Immunohistochemical staining of the lesion demonstrated strong smooth muscle actin expression and focal CD34 expression (figure 2C,D). Proliferative rate on Ki-67 staining was low. The overall morphology and immunoprofile of the lesion were consistent with that of a benign glomus tumour.

Histology confirmed the presence of a benign glomus tumour of the left testis which was completely excised. The patient, reviewed in the outpatient clinic, recovered well from the surgery and was discharged.

DISCUSSION

Glomus tumours are mesenchymal neoplasms that arise from glomus bodies, structures that comprise of an arteriovenous shunt and surrounding connective tissue, that are located in the dermis layer and are responsible for thermoregulatory control. First noted by Wood in 1812, glomus tumours can be categorised into solitary and multiple types.¹⁵ Solitary glomus tumours are most often found in the extremities and characteristically present with a triad of pain, temperature hypersensitivity and tenderness.³ In contrast, multiple-type glomus tumour occurrence can be in extracutaneous locations and is more often recorded as asymptomatic. Overall, glomus tumours make up 1–5% of hand tumours with an estimated 25–75% of these occurring in the subungual region¹⁶ although occurrence in other tissues such as the liver, glans penis, lungs and heart is noted.^{4–8} Glomus tumours are most often reported as benign lesions categorised histologically into glomus tumour proper, glomangioma and glomangiomyoma.¹⁷ Despite being predominantly benign lesions, further investigation should still be a priority in the light of potentially significant local invasion and adjacent structure compression.^{8, 18} Mortality has also been noted in rare cases where lesions metastasise.⁶ Features predisposing to metastasis include multiple-type tumours that are deep in location, of more than 2 cm in size with atypical mitotic figures and a moderate-to-high nuclear grade mitotic activity.¹⁹ There is also a noted risk of spreading when essentially benign glomus tumours are accompanied by sarcomas, as in rare cases of glomangiosarcoma.²⁰ Patterns of metastasis are not particularly well-defined but spread to lymph nodes and blood vessels are the distributions most often reported.⁶

Investigation of a mass arising from the testis is via US in the first instance, with measurement of tumour markers and CT CAP for staging in line with the European Association of Urology (EAU) 2019 recommendations²¹ for management of testicular tumours. These modalities provide some diagnostic information although definitive characterisation of the lesion is obtained post-histological assessment as tumour markers are of limited efficacy in this size of lesion,¹² and although an USS is accurate enough to distinguish testicular from paratesticular lesions, it does not allow for specific differentiation between benign and malignant lesions.^{22, 23} Lesion evolution is monitored over time with US and though there is no specific guidance on the time interval over which repeat imaging should occur, growth has been noted in testicular lesions over both 3-month and 4-month periods, with up to 3 mm enlargement occurring within the 3-month interval.^{24, 25} In this case, US surveillance intervals of 4 weeks and then 3 weeks were pursued. This was undertaken due to the concern that the lesion had shown significant evolution in the 4-week period between the local and first follow-up USS. Though a shorter follow-up interval in comparison to the period

Figure 2 Histopathology (A–D clockwise). (A) H&E stain demonstrating a well-circumscribed and vascular tumour. (B) Round and oval cells with fine granular nuclear chromatin pattern on high magnification. (C) Strong expression of smooth muscle actin on immunohistochemical staining. (D) Focal positivity for CD34 on immunohistochemical staining.

previously noted in the literature, the USS 3 weeks later was to evaluate if further lesional changes had occurred. Furthermore, when considering that small nodules are the origin of any malignant testicular tumour²⁶ and that the doubling time for a germ cell tumour is 10–33 days,²⁷ it seems reasonable to perform an early repeat USS, although we acknowledge the argument for longer intervals between repeat scans.

Such prevalent use of trans-scrotal US has resulted in increased detection of impalpable testicular lesions, often <10 mm in maximal diameter.^{28 29} This is the case with our patient as presentation with right testicular pain resulted in incidental discovery of a left testis lesion measuring approximately 6 mm. Extensive analysis aimed to identify optimal investigation and management of such small, incidentally discovered and unidentified, testicular lesions.¹² This has led to a proposed flow chart on how best to assess and manage patients with such lesions when considering testis-sparing surgery.¹² Particular focus has been given to these cohorts to investigate if incidental discoveries of benign or non-evolving lesions lead to unnecessary radical orchidectomies with patients possibly becoming ‘victims of modern imaging techniques’.¹³ Options for consideration in those with negative tumour markers and small US-identified lesions include radiological monitoring,¹⁴ FSE^{30–33} and partial orchidectomy.

Radiological surveillance is the mainstay of monitoring potentially malignant disease and previous analysis in those with similar-sized lesions showed over 20% ultimately underwent operative management.¹⁴ Only one of the cases proceeding to surgery had malignant disease and the majority were operated on due to the patient’s choice to proceed, even in the presence of non-evolving lesions. This demonstrates the effectiveness of monitoring in avoiding intervention but also highlights the role of the patient’s choice, as demonstrated in our case, with a proportion of patients still opting for invasive management even with no signs of lesion progression. Previous use of surgical sampling techniques prior to radical orchidectomy in such small lesions is also reported as FSE^{30 34 35} and diagnostic biopsies²⁶ were both used. The National Comprehensive Cancer Network indicates that an inguinal biopsy should be considered in an ambiguous intratesticular mass identified on US.³⁶ EAU guidelines state that in cases of undetermined, non-palpable, testicular masses measuring <1 cm on US, FSE is proven to be highly concordant with final histopathology.³⁷ Other work has also noted FSE to be positive in its clinical contributions to diagnosis and operative decision-making.³⁸ However, there is well-reported incidence of false negatives when using FSE^{30 31 33} and it has been stated that FSE may not help avoid unnecessary orchidectomy in routine urological practice in cases of small, impalpable and challenging lesions.¹² More recently, further FSE use has proven very accurate, allowing patients with lesions of approximately 10 mm to be listed appropriately for partial and radical orchidectomies.³² Successful FSE in our case may have avoided radical orchidectomy. Partial orchidectomy was also discussed and offered to the patient but this was not the preferred patient-choice and it may have proved challenging to preserve the testicular parenchyma due to the location of the lesion within the testis. In patients of a different demography for whom fertility was an ongoing consideration, there may have been greater inclination to have testis-sparing surgery or to explore sperm preservation methods prior to orchidectomy.

Ultimately the rationale for offering the patient radical orchidectomy was the concern regarding the growth of the lesion in the absence of a definitive diagnosis, with it possibly representing an evolving neoplasm in the presence of a grossly normal contralateral testis. The patient was fully counselled and

Table 1 Demographics and characteristics of reported cases of testicular glomus tumours

	Tullie <i>et al</i> (this article)	Garg <i>et al</i> ¹¹
Age (years)	47	73
Ethnicity	White, British	NA
Body mass index	24	NA
Tumour size	5.6 mm	1 cm
Tumour location	Central testis	Lower pole
Tumour markers	Normal	Normal

was aware the lesion could prove to be benign. His rationale for not undertaking further monitoring was to avoid repeat appointments or experience potential lesion evolution when he was active, self-employed and had a relatively young family which he was not intending to add to. It was apparent that he was keen to have both a definitive solution and avoid further procedure. Clinical discussion concluded that the preferred option was radical orchidectomy, a decision which the patient was pleased with at follow-up even on identification of a benign glomus tumour. With this in mind, we postulate that the patient would still have chosen to proceed to radical orchidectomy even if FSE or biopsy had proven the lesion to be benign. The unpredictability of benign glomus tumours with potential local invasion should be considered, particularly in the testis where there is such a paucity of cases and hence very little awareness of predicted outcomes. To our knowledge, this is only the second reported case of glomus tumour arising in the testis^{9–11} with a comparison of known factors displayed (table 1).

In the previous reported case of a testicular glomus tumour, as in our case, complete excision was curative with no local recurrence¹¹; the patient made an excellent recovery and was discharged from follow-up.

Learning points

- Investigation of testicular masses should follow current recommendations and includes ultrasound scan, tumour markers and CT staging.
- Glomus tumours may arise in the testis, are a rare cause of testicular neoplasms, and should be considered in the differential diagnosis of small testicular lesions.
- The case highlights the decision-making process around the investigation and management of incidentally discovered, small, testicular lesions, including the potential pitfalls with a lack of clear clinical guidance.
- Management of small, incidentally discovered testicular lesions should consider a range of options, including monitoring, frozen section excision and both partial and radical orchidectomy. The patient should be well-counselled concerning such lesions and have a role in the clinical decision-making process.
- The case highlights how patient attitudes to possible cancer diagnoses can dictate the management course. Such patient decisions can often result in invasive treatment options to eradicate any potential for disease evolution or future recurrence.

Contributors STET was involved in the compilation of the material, collation of the case details, writing of the manuscript and subsequent redrafting. MKQ was the proposer of the concept, involved in the collection of the material, editing and final approval of the manuscript. TK compiled the pathology report, collating and annotating images suitable for publication. SA was the senior author, was involved

in concept proposal, production of images and annotations relevant to the pathology section and gave final approval for the manuscript prior to submission.

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