International Journal of Surgery Case Reports 94 (2022) 107058

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.elsevier.com/locate/ijscr

Case report

# Identifying the uncommon solitary fibrous tumour in a rare location – A case report

Shanthi Periasamy [a,d,*], Anita Mani [a], Graham J. Stewart [b,c], Jacob P. Hampton [b]

[a] Department of Anatomical Pathology, Pathology North, Tamworth Rural Referral Hospital, 168 Johnston street, Tamworth, NSW 2340, Australia
[b] Department of Surgery, Armidale Rural Referral Hospital, 226 Rusden street, Armidale, NSW 2350, Australia
[c] School of Rural Medicine, University of New England, Elm Avenue, Armidale, NSW 2351, Australia
[d] Department of Anatomical Pathology, Pathology North, John Hunter Hospital, NSW 2305, Australia

ARTICLE INFO

*Keywords:*
Case report
Solitary fibrous tumour
Natal cleft
STAT6

ABSTRACT

*Introduction and importance:* The authors describe a case work up of an unusual natal cleft soft tissue tumour which eventually was concluded as solitary fibrous tumour. Solitary fibrous tumour is an uncommon fibroblastic tumour which commonly presents in pleural region and very rarely in extrapleural location such as natal cleft. Accurate identification of this tumour, considering the indeterminate nature of the tumour irrespective of the location, avoided misdiagnosis and played a vital role in follow up management of the patient.

*Case presentation:* 43 year old female patient presented with a painless lump in the natal cleft region for a short duration of 7 months with no other significant medical or surgical or family or psychosocial history. She was not on any regular medications.

*Clinical discussion:* Due to unusual location of the tumour broad range of differential diagnosis including benign and malignant entities were discussed based on histological findings and immunophenotyping. Mammary type myofibroblastoma, glomus tumour, clear cell renal cell carcinoma and clear cell sarcoma of soft tissue were few to note. STAT6 immunohistochemical stain played a crucial role in unravelling this case. Due to gradual increase in size of the lump complete surgical excision was performed with no local recurrence of the tumour.

*Conclusion:* It is important to recognise this rare mesenchymal tumour with intermediate malignant potential occurring at uncommon natal cleft region for which complete surgical enucleation is curative.

## 1. Introduction

Solitary fibrous tumour is a rare fibroblastic soft tissue tumour of intermediate malignant potential. Originally described as a pleural based lesion [1] and most commonly occurring in thoracic cavity [1], it has increasingly been reported at varied extrapulmonary sites like superficial and deep soft tissues [2,3], within visceral organs [2], head and neck [3–5] and meninges [6]. The rare soft tissue locations reported in the literature are back [2,7], buttock [2,4,7,8], perineum [7], groin [7], thigh [4,9] and ischioanal fossa [10]. We report a case of solitary fibrous tumour in an unusual location of natal cleft region which was not reported in literature, however 2 cases were reported in perineal region [7,10]. The challenges of diagnosing this uncommon tumour in the perineal region in a female with broad differentials to consider are discussed. Complete surgical removal was performed in a rural referral hospital.

This work has been reported in line with the SCARE 2020 criteria as per the following article.

Agha RA, Franchi T, Sohrabi C, Mathew G, for the SCARE Group. The SCARE 2020 Guideline: Updating Consensus Surgical CAse REport (SCARE) Guidelines, International Journal of Surgery 2020; 84: 226–230 [11].

## 2. Case presentation

A 43-year-old moderately built female presented with a painless bruise like lump in the natal cleft region of 7 months duration. There was no significant medical or surgical or family or psychosocial history. She was not on any regular medications. Ultrasound detected a 35 mm vascular nodule in the natal cleft subcutaneous tissue plane. Patient was concerned due to gradual increase in size of the lump hence she was referred to general surgeon by her general practitioner. Complete

* Corresponding author at: Department of Anatomical Pathology, Pathology North, John Hunter Hospital, NSW 2305, Australia.
*E-mail address:* shanthi.periasamy@health.nsw.gov.au (S. Periasamy).

https://doi.org/10.1016/j.ijscr.2022.107058
Received 6 February 2022; Received in revised form 5 April 2022; Accepted 5 April 2022
Available online 8 April 2022

surgical enucleation was performed on elective basis by the general surgeon with no postoperative complications.

Macroscopic examination revealed a 30 × 25 × 22 mm globoid, smooth, lobulated and encapsulated soft tissue piece. Cut surface was homogenously light brown to light pink with thin pale fibrous streaks running across forming a vague lobulated appearance (Fig. 1).

Microscopically, the lesion was nodular, well circumscribed and highly vascular with compressed fibrous tissue at the periphery. The lesion consisted of hypocellular and hypercellular areas with the former containing abundant pale stromal collagen and the latter with less intervening stroma (Fig. 2). The tumour demonstrated haphazardly distributed spindle to oval cells arranged around branching and hyalinised variably sized blood vessels. The cells were relatively monomorphic and contained moderate amount of pale eosinophilic to clear cytoplasm with indistinct cell border (Fig. 3). The larger blood vessels imparted a staghorn like appearance. Areas of oedema, hyalinisation and myxoid changes were also evident in the mild chronically inflamed stroma. Mitotic rate was up to 3 per 10 high power field. No necrosis was noted. The tumour abutted the circumscribed resection margin, however infiltration of fibrous pseudocapsule was not identified.

The neoplastic spindle cells demonstrated strong and diffuse nuclear positivity to STAT6 (Fig. 4) and membranous positivity to CD34. Bcl2 showed strong cytoplasmic positivity with perinuclear accentuation and some nuclear staining. In addition, the cells showed moderate positive staining with vimentin, progesterone receptor, CAM 5.2 and neuron-specific enolase. Smooth muscle actin and h-caldesmon showed focal positivity in these cells and around blood vessels. Ki67 showed a slightly higher than expected proliferation index of 10%.

The cells were negative for carbonic anhydrase IX, CD10, S100, HMB45, BRAF V600E, CD31, desmin and inhibin.

The overall features were hence consistent with benign solitary fibrous tumour. Close clinical follow up for an year showed no local recurrence.

## 3. Clinical discussion

Solitary fibrous tumour is a rare mesenchymal neoplasm first described and reported to arise from pleura in 1931 [1]. Subsequently other extra pleural sites were increasingly recognised, especially in the superficial and deep soft tissues like extremities [2], back [2,7], buttock [2,4,7,8], perineum [7], groin [7], thigh [4,9] and ischioanal fossa [10]. Hitherto, extensive literature search showed no report of solitary fibrous

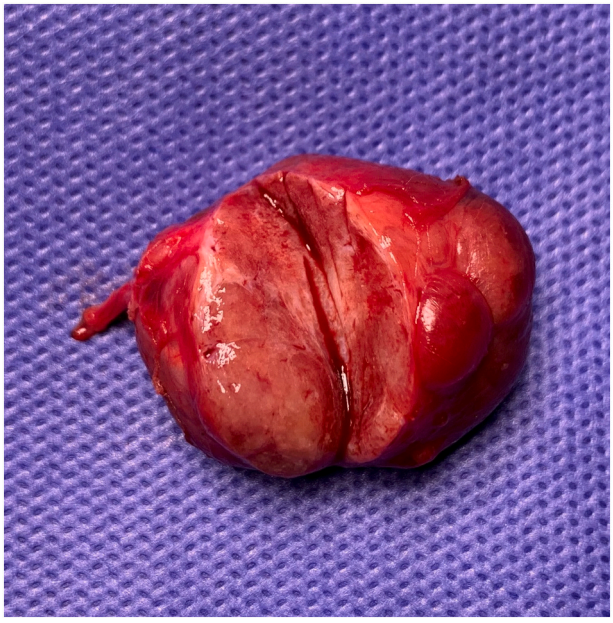

**Fig. 1.** Macroscopy shows smooth, encapsulated tumour with homogenous cut surface.

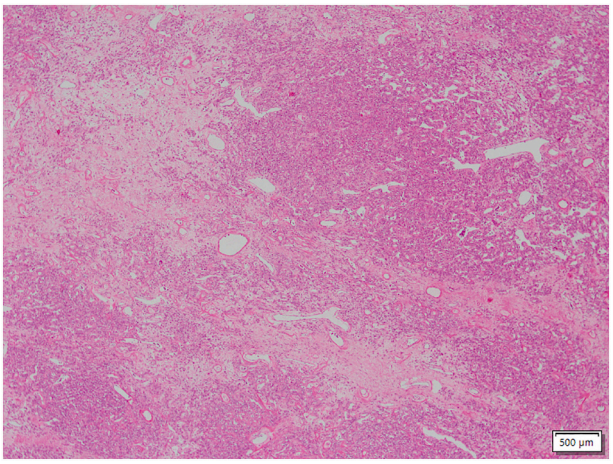


**Fig. 2.** Low power microscopic view of H&E shows hypercellular and hypocellular areas admixed with ectatic blood vessels.

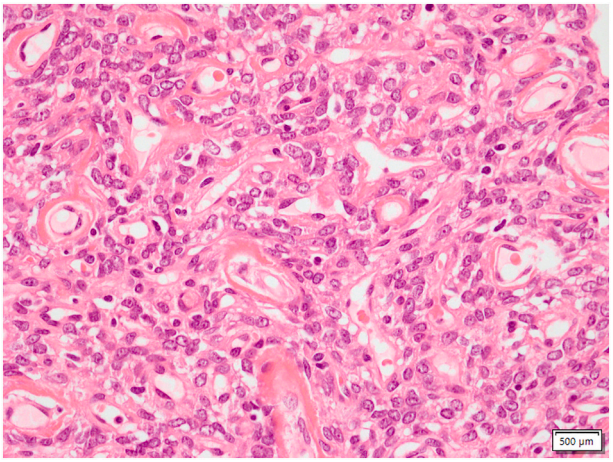


**Fig. 3.** High power microscopic view of H&E shows monotonous spindle to oval cells with indistinct cell border and moderate amount of eosinophilic to clear cytoplasm.

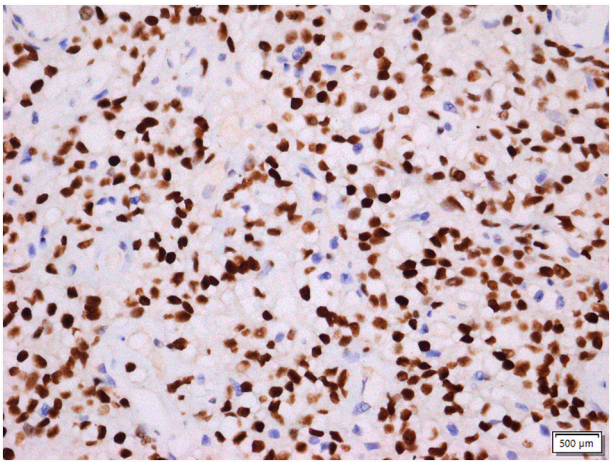


**Fig. 4.** Diffuse and strong nuclear staining with STAT6 immunostaining.

tumour arising particularly from natal cleft though two cases were reported in perineal region [7,10].

Solitary fibrous tumours are slow growing painless masses similar to our case. On rare occasions, solitary fibrous tumour can present with paraneoplastic syndromes, the most described being non-islet cell hypoglycemia, due to tumour production of high molecular weight insulin like growth factor. However, preoperative blood glucose analysis was not performed in our case.

The location of the lesion and some unusual immuno-morphological features posed a diagnostic challenge considering few differentials ranging from benign to malignant tumours. The benign and malignant entities ruled out using immunohistochemistry are discussed here.

Owing to the superficial perineal location in a female, likelihood of mammary type myofibroblastoma, angiomyofibroblastoma and cellular angiofibroma were considered initially. All these tumours share a similar histopathological and immunohistochemical features with minor differences. Mammary type myofibroblastoma, angiomyofibroblastoma and cellular angiofibroma typically present as slow growing painless masses in the pelviperineal region. These tumours are well circumscribed, unencapsulated and characterised by hyper and hypocellular areas of spindle cells in short fascicles which express desmin in a decreasing frequency. Variably sized thin and thick walled, often hyalinised blood vessels and ropey to thin wisps of collagen bundles are interspersed within the tumour. The spindle cells in angiomyofibroblastoma and cellular angiofibroma coexpress vimentin, oestrogen and progesterone receptors, while mammary type myofibroblastoma and cellular angiofibroma show positive staining with CD34 and smooth muscle actin in a differing frequency. Interestingly, in the literature review, solitary fibrous tumour was considered in the differential diagnosis of mammary type myofibroblastoma [12] and cellular angiofibroma [13]. Desmin and other morphological features distinguished both these entities from solitary fibrous tumour respectively.

Due to the perivascular arrangement of tumour cells, glomus tumour, perivascular epithelioid cell tumours (PEComa) and soft tissue hemangioblastoma were contemplated. Glomus tumours classically present in subungual location. However, they were recognised in coccyx by few authors [14] which was debated by others [15]. PEComa is less common in soft tissues. The above two tumours exhibit epithelioid to spindled relatively uniform cells surrounding numerous blood vessels of varying calibre and stain with smooth muscle markers. In addition, glomus tumour expresses type IV collagen and PEComa typically shows melanocytic immunoprofile. Soft tissue hemangioblastoma shows proliferation of stromal cells arranged between numerous small closely packed thin-walled blood vessels which stains with inhibin, neuron-specific enolase, S100, vimentin and CD34. This tumour was considered as a least possibility due to location of the tumour in our case.

The clear cell nature of the tumour imposed metastatic clear cell renal cell carcinoma of low nucleolar grade and clear cell sarcoma of soft tissue in the differential diagnosis. Both tumours are characterised by solid to nested growth pattern separated by thin fibrovascular to fibrous septa. The cellular morphology varies from atypical to bland nuclei with or without prominent nucleoli. Mitotic activity is low in general. While metastatic renal cell carcinoma in perineal region is well documented [16], clear cell sarcoma of soft tissue is not cited in the literature. The earlier tumour is highlighted by carbonic anhydrase IX, Pax8, vimentin, epithelial membrane antigen, CD10, cytokeratin and the latter shows positivity with melanocyte markers. Though our tumour lacked solid to nested growth pattern, the cellular features raised a possibility of these tumours, however immunohistochemistry findings did not support the same.

Our case showed diffuse and strong nuclear immunoreactivity for STAT6 which has consistently been reported in almost all of solitary fibrous tumour cases [17]. Strong and diffuse expression of CD34 and bcl2 were also demonstrated in various studies [7,18]. Aberrant cytokeratin expression can be seen, especially in malignant solitary fibrous tumours [19]. Interestingly, patchy cytokeratin expression was observed in our case although no malignant features were identified as described in few studies [4] such as tumour size >4 cm, high mitotic count >4 mitoses per 10 high power fields, high cellularity, nuclear pleomorphism and foci of haemorrhage and necrosis.

Distant and local recurrences can occur in 10–30% of cases. Due to recurrences even in solitary fibrous tumours lacking atypical features, a three-tier risk stratification model by Demicca et al. [20] is used to stratify the patients accurately for close follow up. Our patient fell under low-risk group and was disease free in the 12-month close follow up.

## 4. Conclusion

Due to the rarity of solitary fibrous tumour in extrapulmonary sites, missed or misdiagnosis is a potential risk. Irrespective of their location, diagnosis of solitary fibrous tumour should be considered in soft tissue spindle cell neoplasm associated with variable sized blood vessels and collagenous stroma. Due to its intermediate malignant potential, risk stratification system should be included in routine histopathological reporting to determine the recurrence risk and follow up time period, which will aid the clinicians in the management.

## Provenance and peer review

Not commissioned, externally peer-reviewed.

## Sources of funding

None.

## Ethical approval

Not applicable.

## Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

## Author contribution

**S Periasamy**: Data curation, manuscript writing, reviewing, editing, visualisation; **A Mani**: Conceptualisation, supervision; **G Stewart and J Hampton**: patient history, surgical removal of specimen and macroscopic image.

## Research registration

Not applicable.

## Guarantor

Shanthi Periasamy.

## Declaration of competing interest

None.

## References

[1] P. Klemperer, B.R. Coleman, Primary neoplasms of the pleura. A report of five cases, Am. J. Ind. Med. 22 (1) (1992) 1–31, https://doi.org/10.1002/ajim.4700220103.

[2] O.J. Wignall, E.C. Moskovic, K. Thway, J.M. Thomas, Solitary fibrous tumors of the soft tissues: review of the imaging and clinical features with histopathologic

correlation, AJR Am. J. Roentgenol. 195 (2010) W55–W62. https://www.ajronline.org/doi/10.2214/AJR.09.3379.
[3] M. Fukunaga, H. Naganuma, T. Nikaido, T. Harada, S. Ushigome, Extrapleural solitary fibrous tumor: a report of seven cases, Mod. Pathol. 10 (1997) 443–450.
[4] G.P. Nielsen, J.X. O'Connell, G.R. Dickersin, A.E. Rosenberg, Solitary fibrous tumor of soft tissue: a report of 15 cases, including 5 malignant examples with light microscopic, immunohistochemical, and ultrastructural data, Mod. Pathol. 10 (1997) 1028–1037.
[5] D.M. Dorfman, K. To, G.R. Dickersin, A.E. Rosenberg, B.Z. Pilch, Solitary fibrous tumor of the orbit, Am. J. Surg. Pathol. 18 (3) (1994) 281–287, https://doi.org/10.1097/00000478-199403000-00008.
[6] S.S. Carneiro, B.W. Scheithauer, A.G. Nascimento, T. Hirose, D.H. Davis, Solitary fibrous tumor of the meninges: a lesion distinct from fibrous meningioma. A clinicopathologic and immunohistochemical study, Am. J. Clin. Pathol. 106 (2) (1996) 217–224, https://doi.org/10.1093/ajcp/106.2.217.
[7] S. Suster, A.G. Nascimento, M. Miettinen, J.Z. Sickel, C.A. Moran, Solitary fibrous tumors of soft tissue. A clinicopathologic and immunohistochemical study of 12 cases, Am. J. Surg. Pathol. 19 (11) (1995) 1257–1266, https://doi.org/10.1097/00000478-199511000-00005.
[8] V. Vigorita, M.B. Zoccali, S. Rausei, et al., Giant solitary fibrous tumor of the buttock, ACG Case Rep. J. 3 (2) (2016) 139–140, https://doi.org/10.14309/crj.2016.26. Published 2016 Jan 20.
[9] M. Martorell, A. Pérez-Vallés, F. Gozalbo, J.A. Garcia-Garcia, J. Gutierrez, J. Gaona, Solitary fibrous tumor of the thigh with epithelioid features: a case report, Diagn. Pathol. 2 (2007) 19, https://doi.org/10.1186/1746-1596-2-19. Published 2007 Jun 18.
[10] A. Bhat, L.J. Layfield, S.O. Tewari, A.H. Gaballah, R. Davis, Z. Wu, Solitary fibrous tumor of the ischioanal fossa-a multidisciplinary approach to management with radiologic-pathologic correlation, Radiol. Case Rep. 13 (2) (2018) 468–474, https://doi.org/10.1016/j.radcr.2018.01.030. Published 2018 Mar 2.
[11] R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, for the SCARE Group, The SCARE 2020 guideline: updating consensus Surgical CAse REport (SCARE) guidelines, Int. J. Surg. 84 (2020) 226–230.
[12] S. An, J.S. Song, S. Park, J.W. Lee, K.J. Cho, Mammary-type myofibroblastoma: a report of two cases, J. Pathol. Transl. Med. 50 (5) (2016) 385–389, https://doi.org/10.4132/jptm.2016.03.26.
[13] U. Flucke, J.H. van Krieken, T. Mentzel, Cellular angiofibroma: analysis of 25 cases emphasizing its relationship to spindle cell lipoma and mammary-type myofibroblastoma, Mod. Pathol. 24 (1) (2011 Jan) 82–89, https://doi.org/10.1038/modpathol.2010.170.
[14] L. Duncan, J. Halverson, K. DeSchryver-Kecskemeti, Glomus tumor of the coccyx. A curable cause of coccygodynia, Arch. Pathol. Lab. Med. 115 (1) (1991) 78–80.
[15] Z. Gatalica, Lucio E.T. Liqiang, M. Miettinen, Glomus coccygeum in surgical pathology specimens: small troublemaker, Arch. Pathol. Lab. Med. 123 (10) (1999) 905–908. http://search.proquest.com.acs.hcn.com.au/scholarly-journals/glomus-coccygeum-surgical-pathology-specimens/docview/211956555/se-2?accountid=130851.
[16] G.W. Mendese, P.J. Ayvazian, C. Li, Renal cell carcinoma presenting as a perineal mass: case report and review of the literature, Urology 67 (4) (2006), https://doi.org/10.1016/j.urology.2005.10.047, 847.e1-847.e8472.
[17] L. Doyle, M. Vivero, C. Fletcher, F. Mertens, J. Hornick, Nuclear expression of STAT6 distinguishes solitary fibrous tumor from histologic mimics, Mod. Pathol. 27 (2014) 390–395, https://doi.org/10.1038/modpathol.2013.164.
[18] T. Hasegawa, T. Hirose, K. Seki, P. Yang, T. Sano, Solitary fibrous tumor of the soft tissue. An immunohistochemical and ultrastructural study, Am. J. Clin. Pathol. 106 (3) (1996) 325–331, https://doi.org/10.1093/ajcp/106.3.325.
[19] K. Akaike, A. Kurisaki-Arakawa, K. Hara, et al., Distinct clinicopathological features of NAB2-STAT6 fusion gene variants in solitary fibrous tumor with emphasis on the acquisition of highly malignant potential, Hum. Pathol. 46 (3) (2015) 347–356, https://doi.org/10.1016/j.humpath.2014.11.018.
[20] E.G. Demicco, M.J. Wagner, R.G. Maki, et al., Risk assessment in solitary fibrous tumors: validation and refinement of a risk stratification model, Mod. Pathol. 30 (10) (2017) 1433–1442, https://doi.org/10.1038/modpathol.2017.54.