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Undifferentiated round cell and spindle cell sarcomas: the current paradigm

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Introduction: The classification of soft tissue and bone neoplasms is experiencing a transformation as methods to detect gene fusions expand. Next-generation sequencing panels, anchored multiplex polymerase chain reaction systems and comprehensive RNA sequencing have all contributed to new histological categorisation. The EWSR1-NFATC2 is one such novel translocation found in a round cell sarcomas. These sarcoma sit within the WHO group of undifferentiated round and spindle cell sarcomas. They presents as either a primary bone or soft tissue tumour and exhibits distinctive histopathologic features.

Methods: This is a case study of a soft tissue mesenchymal tumour with the recently described gene fusion EWSR1-NFATC2. The case is described, with 3-month outcomes and considered in the most recent literature on this sarcoma subset (26 cases).

Results: This case study prompted a look at the current NHS paradigm for these emerging histopathologically distinctive sarcoma subgroups.

At present the expertise with the sarcoma MDT, along with second opinion from other national MDTs is the format for subtype specific sarcoma management, informed by recent literature.

Conclusion: The EWSR1-NFATC2 fusion round cell sarcoma tumours are an example of a very recently emerged sarcoma subset which display oncological variance to the wider Ewing's family. The novel literature guided the Southmead and Royal Marsden MDT opinions to proceed straight to surgical excision.

The Authors recommend a national prospective database for rare sarcoma subsets, particularly those that appear to exhibit distinctive clinicopathology characteristics. The format could be through the RSTN collaborative, secure anonymised data collection can be collected through REDCap.