Royal Australasian College of Surgeons

Massive myxopapillary ependymoma of the gluteal cleft

A 19-year-old male presented with a 2-year history of a progressively enlarging sacrococcygeal mass. Despite the significant size of the mass resulting in disfigurement and difficulty maintaining hygiene, he presented after the development of ulceration of the overlying skin and associated bleeding (Fig. 1). He denied any associated bladder or bowel changes, and had a normal lower limb neurological examination. He had no significant past medical history with no regular medications. Further investigation included a pelvic Magnetic Resonance Image (MRI) (Fig. 2) which showed an 11.4 × 9.9 × 10.3 cm lesion arising from the coccyx extending into the subcutaneous soft tissues of the gluteal cleft, characterized by multiple internal septations, marked T2 hyperintensity and avid Gadolinium enhancement. Histological examination of an image-guided core biopsy was suggestive of a myxopapillary ependymoma.

A resection of the sacrococcygeal tumour was performed under general anaesthesia in the prone position. Macroscopically clear margins were achieved with en bloc resection of gluteal muscle, fascia and the coccyx. The wound was closed primarily with interrupted sutures and a suction drain to the resection cavity.

Microscopic pathological assessment of the tumour revealed cuboidal tumour cells radially arranged in a papillary manner around vascularized stromal cores, separated by abundant myxoid matrix. The tumour extended beyond the deep fascia and into the marrow of the coccyx. Lymphovascular and perineural invasion were present. The immunocytochemical assessment demonstrated expression of the glial fibrillary acidic protein, but was negative for CK5/6, cytokeratin and SOX10, consistent with the preoperative diagnosis of myxopapillary ependymoma and differentiating the lesion from the more common chordoma. Resection margins were microscopically clear. The patient had an uncomplicated

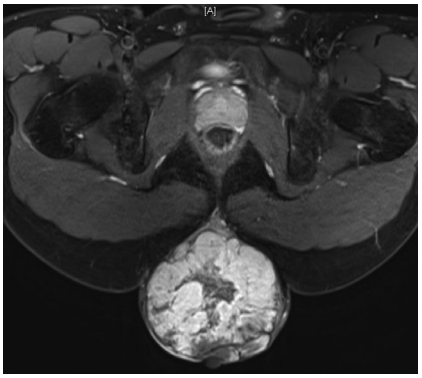

Fig. 2. Axial MRI images of sacral region demonstrating an exophytic mass with gadolinium contrast enhancement on T1.

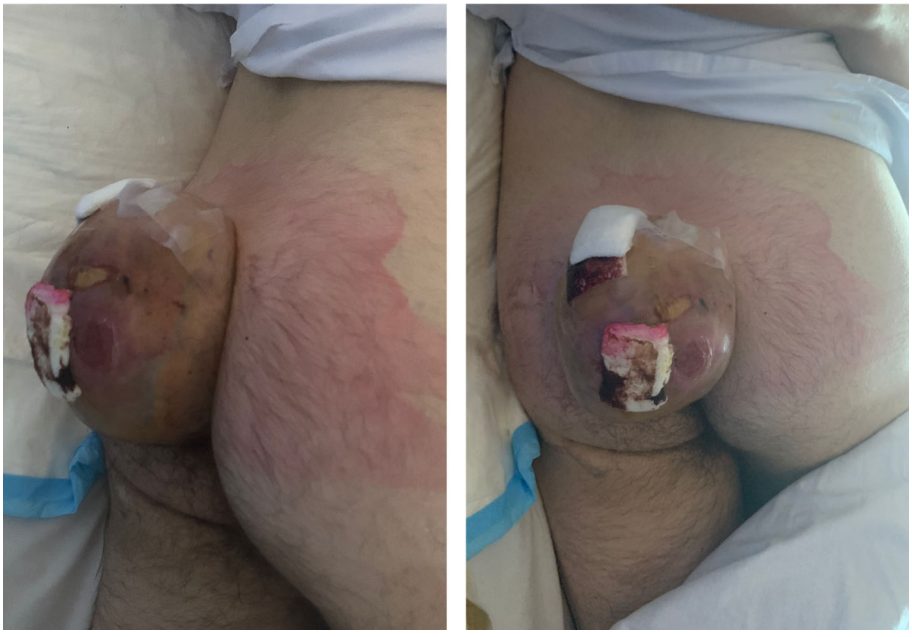

Fig. 1. Clinical photograph of massive soft tissue lesion of the gluteal cleft with ulceration of the overlying skin (dressings applied for haemostasis prior to referral to our facility).

post-operative recovery and was discharged home on Day 7. At 1-year follow-up, he remains well, with no clinical or radiological evidence of recurrent disease.

A myxopapillary ependymoma is a slow-growing histological variant of ependymoma that commonly originate within the filum terminale or conus medullaris in the lumbosacral region.[1,2] Virchow[3] first identified ependymal tumours in 1865; however, it was not until 1935 when Kernohan and Fletcher-Kerohan[4] sub-classified a selection of these tumours as myxopapillary ependymomas. They are currently classified according to the World Health Organization Classification of Tumours of the Nervous System as Grade 1 gliomas.[5] Myxopapillary ependymomas are rare, with an incidence of 1 per million person-years.[6] The average age of diagnosis of these tumours is 36, however, tumours found in paediatric patients are generally more aggressive and are more likely to metastasise.[7] Grossly, these tumours are usually small encapsulated masses with gelatinous interiors. Microscopically, the characteristic features of the tumours are a pseudopapillary pattern of cuboidal tumour cells around a vascular stromal core that undergoes mucinous degeneration.[8] Though considered to have indolent biological behaviour, up to 31.7% of patients will experience disease recurrence. Of these patients, most experience local recurrence (84.5%), however over one-third of patients have non-local disease involving the distant spine and/or brain.[8]

Surgical resection is the mainstay treatment modality for myxopapillary ependymoma. The role of adjuvant radiotherapy is controversial, but may improve local recurrence, progression-free and overall survival in younger patients, particularly in the setting of subtotal resection.[8,9] Younger age, subtotal or piecemeal resection and larger tumour size (>2 cm) confer poorer prognosis. The prognostic significance of massive tumour size such as in the case presented here remains uncertain due to its rarity, and close clinical and radiological surveillance is warranted.

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Author contributions

BJ completed the initial draft. SF, DD, MN & PC were involved in the drafting and editing of the manuscript. All authors viewed the manuscript before submission.

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