

Pleomorphic dermal sarcoma: a case report

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ABSTRACT

A pleomorphic dermal sarcoma (PDS) is considered a rare and poorly understood neoplasm of the skin.¹ This has features similar to atypical fibroxanthoma (AFX).¹ As well as the baseline similar characteristics to an AFX, a PDS also exhibits features of an infiltrative growth pattern which allows invasion of the subcutis, tumour necrosis and the possibility of lymphovascular invasion.² These are more aggressive features than the more extensively reported AFX. There is a degree of metastatic potential associated with a PDS and hence early diagnosis and management with the appropriate multi-disciplinary team (MDT) is important to optimise the clinical prognosis. This case report will discuss a relatively rare diagnosis of a patient presenting with a PDS on the scalp and his management within the Oral and Maxillofacial Surgical (OMFS) Department, including consideration of his overall palliative care. Here, we will discuss the social, medical and physical implications of this diagnosis as well as considering the overall determinants of his quality of life (QoL).

BACKGROUND

A 78-year-old male Caucasian patient presented initially to the OMFS department in 2018 with a squamous cell carcinoma (SCC) on the left parietal scalp. This was excised completely under a local anaesthetic (LA) procedure. This patient's medical history included atrial fibrillation (AF), an aortic valve replacement (AVR) in 2007, in remission for colonic cancer initially resection in 2006 and was borderline diabetic. Following the SCC removal on the scalp, the patient remained under review within the department, later presenting in April 2021 with actinic keratosis with severe dysplasia to the skin at the left vertex. Further histopathological investigation of this

site identified the PDS, which now extended to the full frontal and vertex aspects of the patient's scalp. A computerised tomography (CT) scan reported a locally advanced tumour on the scalp with increased bone destruction and penetration of the tumour into the cranial cavity (see figures 1 and 2). This had direct communication with the anterior aspect of the superior sagittal sinus but no thrombosis and no distant metastasis noted. An MDT meeting conducted in January 2022 suggested supportive patient care with no formal surgical intervention.

DISCUSSION

There are various treatment options and methods regarding lesions and neoplasms of the skin, particularly in the head and neck region. These options include: surgical excision; cryotherapy; topical cytostatic cream; photodynamic therapy; radiotherapy; and electrochemotherapy.³ In this patient's case, radiotherapy was not possible due his compromised renal function caused by previous radiotherapy for colonic malignancy.

As the patient was on a palliative care pathway, it was decided that regular 'debulking of the tumour on the scalp' would be appropriate for managing the size and haemostatic control of the tumour (see figures 3 and 4). Surgical debulking, also known as cytoreductive surgery,⁴ of tumours is where a surgically incurable malignant neoplasm is partially removed without complete curative intent.⁵ This treatment was carried out under local anaesthetic. Due to the irregular size and rapid growth of the tumour, the patient regularly experienced repeated issues of bleeding due to the highly vascularised site. In this patient, the act of debulking assisted with haemostasis of the local site, reduced iron deficiency anaemia and fatigue due to blood loss, as well as cohesively working alongside

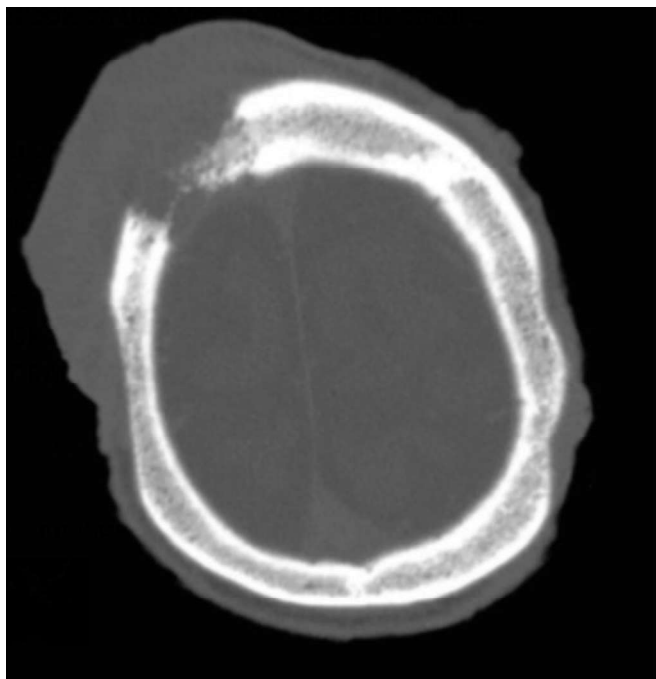


Figure 1: CT scan showing destruction of the parietal bone.

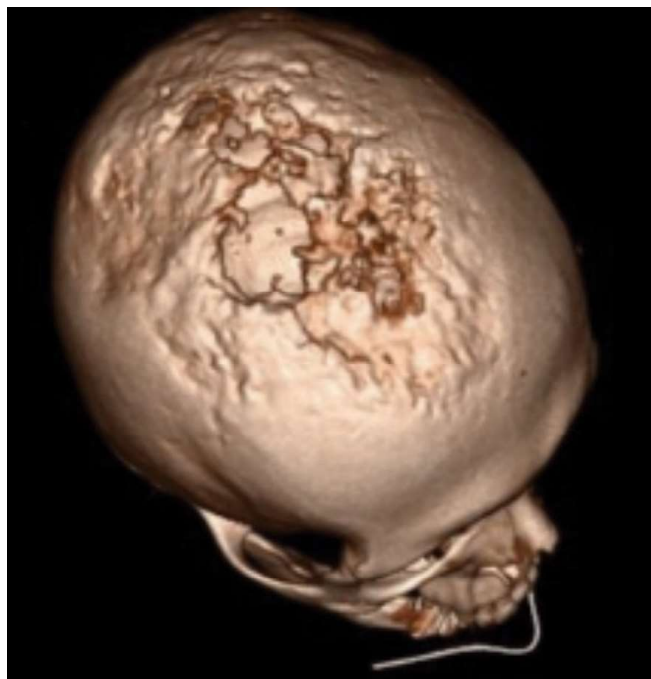


Figure 2: 3D CT scan showing the extent of the PDS destruction of the skull.

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Figure 3: pre-debulking surgical procedure (October 2022).

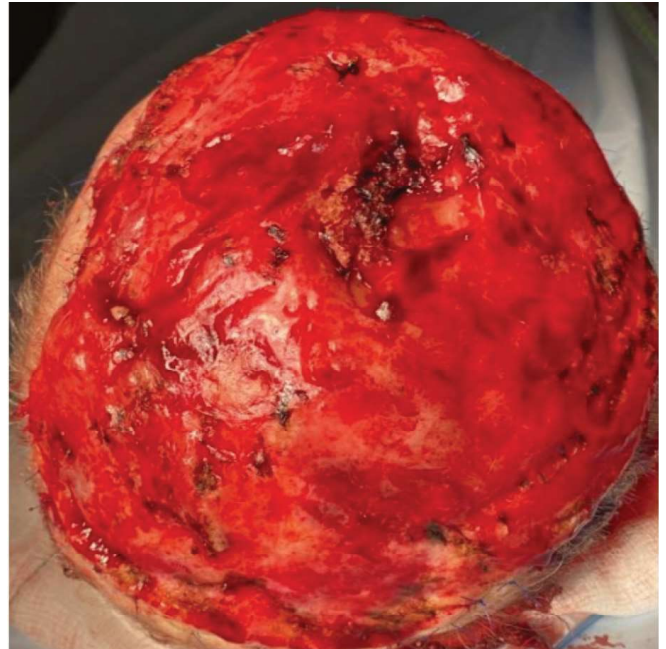


Figure 4: post-debulking surgical procedure (October 2022).

the district general nurses (DGNs) regarding regular wound reviews and dressing changes. The advantages to surgically debulking tumours include preventing deterioration of a patient's QoL.⁴ The act of debulking was a factor to consider when regarding the psychosocial factors for both patients and their carers.⁶ In this case the patient's elderly wife was the sole carer, who had to manage the patient's erratic behaviour, periods of memory loss, confusion and aggression, as well as ensure that they attended every hospital appointment.

Health-related diagnosis and consideration for the patient's QoL is integral to the treatment planning process.⁷ The World Health Organization (WHO) defines QoL as an "individual's perception of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations, standards and concerns".⁸ A further term 'health-related quality of life' is specific to a health care diagnosis. This allows healthcare professions to target their assessment of the impact of the disease and its management with the physical, psychological and social aspects of the patient's needs.⁹ A study in 2002 reviewed the relationship between patient-carer with a palliative diagnosis and concluded the significant importance of the health-care professionals providing good-quality information and management options which impacted the patient's overall approach and understanding of their diagnosis.⁶ In this case, autonomous and informed consent for surgical debulking was gained and discussed with the patient and his carer wife.

Due to the aggressive nature of the cancer, the tumour was debulked several times, the first of which was completed on 17th March 2022. The images show the cancer at the third debulking which was completed on 23rd September 2022. During this time the area had grown substantially and there were recurrent issues of bleeding from the highly vascular tumour. Prior to the third debulking, there was discussion of stopping the patient's Warfarin due to the substantial blood loss the patient was sustaining. By doing so, this would make the patient at a greater risk of a stroke. After discussion with the patient and his family, a DNA CPR (do not attempt

cardiopulmonary resuscitation) was put in place.

The debulking also helped to reduce the dressing times and frequency of dressing changes for the patient and reduced the pain from these appointments. The PDS had grown significantly which was putting pressure on the scalp and brain especially when dressings were changed, which was twice a week.

By choosing to regularly debulk the tumour rather than monitor it meant the patient spent less time in hospital; ultimately reducing stress for the patient and his wife. This also provided a considerable cost-cutting benefit to the NHS. Keeping the bulk of the tumour as minimal as possible reduced the chances of the patient experiencing an uncontrolled bleed and having to then be admitted into hospital. 2017 figures suggest that the cost of just one night in a UK hospital costs £589.59 and an outpatient visit £201.00.¹⁰ These costs were avoided by reviewing the patient twice a week in the maxillofacial department's dressings clinic, where the dressings were changed by an experienced nurse. This ensured that the tumour was being cared for effectively, reducing the chances of the patient developing a healthcare acquired infection and prevented the need for him to seek care from the emergency department in the hospital. However, twice-weekly dressing changes still incur a cost, although nowhere near amounting to that of theatre sessions and in-patient stays, remaining ultimately the most cost-effective way of managing the patient. Interestingly a study conducted in 2017 found that £8.3 billion was spent on wound management, of which £5.6 billion was spent on non-healing wounds.¹¹ Whilst still contributing to the financial burden on the NHS via regular dressing changes, tumour debulking was a financially viable option and more importantly greatly improved the QoL for this patient.

CONCLUSION

PDS is an extremely aggressive sarcoma and must be detected and treated early. When a patient cannot receive treatment for an actively growing tumour, palliative debulking

is an excellent way of improving QoL. Making patients as comfortable as possible should be the main priority for any clinician and certainly goes hand in hand with the oath to 'do no harm'. Tumour debulking ensures control over tumours and reduces costs to the NHS overall by limiting secondary complications that can otherwise arise from fast-growing, highly vascular tumours.

CONFLICT OF INTEREST STATEMENT

All authors declare there are no conflicts of interest. This article has not been published elsewhere. Consent for photographs has been obtained.

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