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CASE REPORT

Big Bad BCCs: Craniofacial resection and reconstruction for atypical basal cell carcinomata

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Summary Basal cell carcinoma (BCC) is the most common malignancy of the body and most frequently occurs in the head and neck. 'Problematic aggressive' BCCs are either frequently recurrent, often after histologically confirmed clearance, or are 'giant/horrifying' and invasive of critical anatomy. Three patients that illustrate different clinical features and outcomes of 'problematic aggressive' craniofacial BCCs are reported that required craniofacial resection and reconstruction with microsurgical free flap cover, and a re-evaluation of our care pathway for these uncommon presentations.

Basal cell carcinoma (BCC) is the most common malignancy of the body with 114 cases per 100 000 population in the UK each year.[1] BCCs occur mainly in sun damaged skin of fair skinned individuals later in life and are more common in men. Seventy percent of these tumours occur in the head and neck.[2] BCC originates from pluripotent cells of the epidermis and hair follicles, and is usually indolent with low metastatic rate.[3] A rare subset of BCCs behaves aggressively, frequently recurring despite clear surgical margins.[4] A second clinical subset of BCC tumours can be greatly problematic in the craniofacial complex. These are 'giant',[5] or 'horrifying'[6] basal cell carcinomas, and are TNM stage T3 carcinomas (Table 1) defined as greater than 5 cm at greatest diameter and comprising 0.5 percent of BCCs.[3] Radical resection is required but is often complicated by the invasion of important structures. The size of the primary tumour correlates to the risk of metastasis.[7,8] Metastasis of BCC is rare and reported as 0.0028% in a large Australian study[9] with a mean survival of ten months after the diagnosis of distant spread.[10] Factors that may play a role in the development of giant BCC or the onset of aggressive malignant behaviour include tumour factors, the

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Table 1 AJCC TNM Staging of Basal Cell Carcinoma.

T1:	The tumour is 2 centimetres (cm) at greatest diameter or less
T2:	Tumour 2—5 cm at greatest diameter
T3:	Tumor is larger than 5 cm at greatest diameter
T4:	Tumor of any size that invades deeply into muscle, cartilage, or bone
Nx:	Nearby lymph nodes cannot be assessed
N0:	No spread to nearby lymph nodes
N1:	Spread to nearby lymph nodes
Mx:	Presence of distant metastasis cannot be assessed
M0:	No distant metastasis
M1:	Distant metastasis present

co-morbidity of the patient, previous treatment modalities, and the delay in presentation. We report three cases of 'problematic aggressive' BCCs that illustrate different aspects of the clinical behaviour of these tumours, and that required a radical surgical strategy comprising craniofacial, microsurgical and alloplastic reconstruction.

Case reports

Case 1 - Giant Fronto-orbital BCC

A sixty-four year old retired office manager presented with a scalp ulcer following a curling tong burn to the scalp a year earlier. The painless ulcer with indistinct margins occupied the fronto-parietal scalp and extended onto the ipsilateral ear, the pre-tarsal skin and orbicularis oculi of the right upper eyelid, and across the nasion to the contralateral eyebrow and back to the vertex. Assessment within the multidisciplinary team (MDT) environment included quadrant biopsies and CT scans which confirmed a diagnosis of morphoeic basal cell carcinoma with transcranial invasion (Figure 1).

The scalp component of the tumour was first resected from the calvarium with a minimum 3 cm margin, which was then itself removed as a separate craniectomy specimen with a 2 cm gross margin. A dural resection of tumour was undertaken and the defect closed with a pericranial graft, sealed with fibrin glue (Tisseel, Baxter, IL). Attention was then turned to the orbitopalpebral component of the disease. The soft tissue component of the tumour was removed, continuing the skin incision from scalp to upper eyelid, glabella and back to the scalp across the midline. Deeply, periosteum was elevated from the frontozygomatic process, exposing the supraorbital margin above and the deep temporal fascia posteriorly, whilst elevating the skin and orbicularis muscle together from the tarsus, levator mechanism and septum of the upper lid. The cutaneous tumour resection margin was less than 1 cm at the lid margin. The sensory nerves of the forehead entered the deep surface of the tumour, and following positive frozen section analysis of the supraorbital nerve at the foramen a segmental resection of the supraorbital foramen and nerve was performed. Cranioplasty was achieved using titanium mesh for the calvarium, and split calvarial bone to reconstitute the supraorbital margin. Soft tissue cover was achieved with a free latissimus dorsi muscle flap, harvested by a second surgical team via a mid-axillary incision. This was anastomosed to the facial vessels and a split thickness skin graft applied to achieve skin cover.

Following the procedure, the patient required evacuation of a haematoma from the right temple deep to the flap. Hospital stay was 12 days and the post-operative course was otherwise unremarkable. There is no evidence of Contoured reconstruction at four years follow up post-operatively (Figure 2)

Case 2 - Giant Fronto-temporo-facial BCC

A seventy-four year old retired office worker was referred with a giant painless ulcerating BCC of the left fronto-parieto-vertex region, extending through the left ear to the mastoid area and petrous temporal bone, as well as *via* the temple and lateral canthus into the left parotido-masseteric area. The tumour was clinically fixed to the fronto-zygomatic process, left temple, vertex and left masseter. The tumour had been present for ten years but grown rapidly over the preceding two months. On the contralateral side there was a further BCC replacing the right post-auricular sulcus. Dual modality imaging demonstrated dural involvement and invasion of the lateral calvarium, greater wing of the sphenoid, and temporal bone, but the middle ear was spared (Figure 3). Pre-operative chest radiographs were unremarkable. Pre-operative biopsies confirmed infiltrative basal cell carcinoma.

Resection was planned in three components: cranio-temporal, orbito-sphenoid, and parotido-masseteric (Figure 4), to achieve a 'compartmentalised *en bloc*' resection. The cranio-temporal component was undertaken first, with soft tissue tumour and infiltrated craniectomy performed *en bloc* with incisions/craniotomy through tumour anteriorly (through the supraorbital/forehead junction) and laterally (through the temporal bone). A 3 cm tumour resection margin was observed medially and posteriorly. Resection of underlying involved dura (with a 1—2 cm margin) resulted in exposure of the underlying meninges and brain. The orbito-sphenoid tumour component was then approached by skin incisions continued through the lateral third of the upper eyelid to the lateral canthus down into the facial part of the resection. The soft tissue tumour margins in the upper lid were maximally 1 cm. The orbito-sphenoid osteotomy included the lateral third of the supraorbital margin and upper fronto-zygomatic process, and the lateral third of the posterior wall of the orbit (greater wing of sphenoid), to meet the anterior part of the temporal osteotomy of the first component resection and expose the underlying periorbita/dura. Finally, the parotido-masseteric resection involved en bloc resection of the pinna, radical parotidectomy with sacrifice of the facial nerve, resection of the zygomatic arch, temporalis muscle and part of the masseter.

This dura was repaired with a pericranial graft and sealed with fibrin glue (Tisseel, Baxter, IL) The zygomatico-frontal process, supraorbital bar, and lateral orbital wall were reconstituted using moulded methylmethacrylate to achieve a natural contour. A titanium mesh cranioplasty was fixed to the calvarium and methylmethacrylate construct, and soft tissue cover was achieved using a free latissimus dorsi muscle