British Journal of Plastic Surgery (1987), **40**, 528–531

Multiple metastases from basal cell naevus syndrome

P. A. WINKLER and B. GUYURON

Division of Plastic Surgery, Mount Sinai Medical Center, Cleveland, Ohio, USA

Summary—A patient with basal cell naevus syndrome (Gorlin-Goltz Syndrome) is presented. Of note in the case was the extensive symmetrical tumour invasion of both external auditory canals requiring bilateral radical resection. The patient expired 14 months later, at which time the autopsy revealed widespread metastases to the pleura, diaphragm, pericardium, epicardium and myocardium. Although lung metastases have been reported in this syndrome, no cases have been reported of metastases to these sites.

The basal cell naevus syndrome (Gorlin-Goltz Syndrome) has come to be a well-known entity since it was first described by Jarish and White in 1894 (Jarish, 1894; White, 1894). Indeed, features of the syndrome have been found in skeletons excavated in lower Egypt dating back to the Dynastic Period (Satinoff and Wells, 1969). Gorlin and Goltz, in 1960, tabulated all the cases to date and suggested a well-defined symptom complex (Gorlin and Goltz, 1960). Their initial tripartite syndrome of basal cell carcinomas (BCC), jaw cysts and skeletal anomalies has since been expanded to include almost forty different anomalies. Excellent review articles have been published recently describing the various findings in patients with this disease, and numerous new cases have been documented (Olson *et al*, 1981; Fitzpatrick and Thompson, 1982). A recent article described the potentially aggressive nature of this syndrome in two cases (de la Plaza *et al.*, 1983). We present here a case of basal cell naevus syndrome with extensive bilateral symmetrical tumour invasion of the external auditory canals and cranial base, in addition to pleural, diaphragmatic, epicardial and pericardial metastases.

Case report

This 48-year-old white male was originally diagnosed as having basal cell naevus syndrome in 1961. He was noted to have multiple basal cell carcinomas of the forehead and a right mandibular cyst. Other syndrome characteristics noted were: hypertelorism, congenital blindness of the right eye, frontal bossing, right-sided facial palsy, curvature of the spine and calcification of the falx cerebri. Family history was negative for BCC.

Between 1972 and 1978, this patient underwent 25 procedures for removal of skin lesions as well as a craniotomy for frontal bone involvement with latissimus dorsi free flap reconstruction. This was followed by radiotherapy (5000 rads) to the forehead. Later in 1978, a left preauricular morphea type BCC lesion appeared and a superficial right parotidectomy with facial nerve biopsy also proved positive for BCC without evidence of squamous differentiation. The preauricular lesions were excised and Cobalt therapy added. In 1981, however, involvement of the left external auditory canal and temporomandibular joint was noted. Two courses of Bleomycin (totalling 200 units) proved ineffective, and Cis-Platinum therapy was started along with Velban therapy and intra-arterial 5-FU. It should be noted that each excision of a new lesion was felt to be adequate at the time of surgery, and that each recurrent lesion was felt to be local spread of a new lesion (Fig. 1).

In December 1983, the patient was admitted to our unit with extensive, bilateral BCC of the face with cranial base and mandibular invasion (Fig. 2A, B). Extensive ulceration was noted in the external auditory canals with decreased hearing on the right side and total hearing loss on the left. Cranial nerve VIII showed decreased function on the right and none on the left. Chest X-ray revealed no metastases. A radical resection was performed which included the right ear, the zygoma, the temporomandibular joint, the muscles of mastication and the contents of the right infra-temporal fossa. A partial right mandibulectomy was also performed with a total right parotidectomy. Reconstruction was achieved with a right-sided pectoralis musculocutaneous island flap and a right postauricular flap. All margins were reported to be free of tumour. The patient did well and was discharged in 1 week. He was subsequently readmitted the following month and underwent an equally radical left-sided resection. Exploration of the foramina ovale and spinosum showed microscopic involvement of V_2 nerve and middle meningeal artery. A pectoralis major musculocutaneous flap was used to provide adequate tissue coverage to the left side. Postoperatively, the patient received radiotherapy to the cranium. After the initial

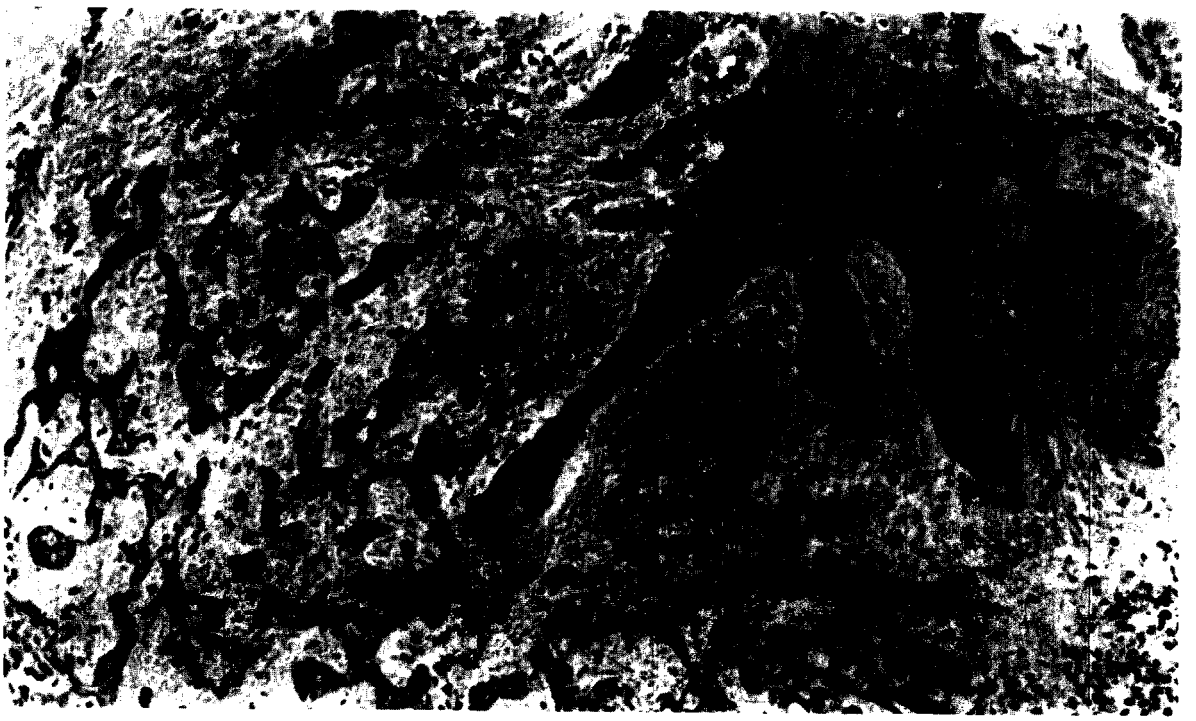

Fig. 1

Figure 1—Primary skin lesion exhibiting a morphea-like proliferating pattern from the origin of the tumour at the epidermis. (10×).

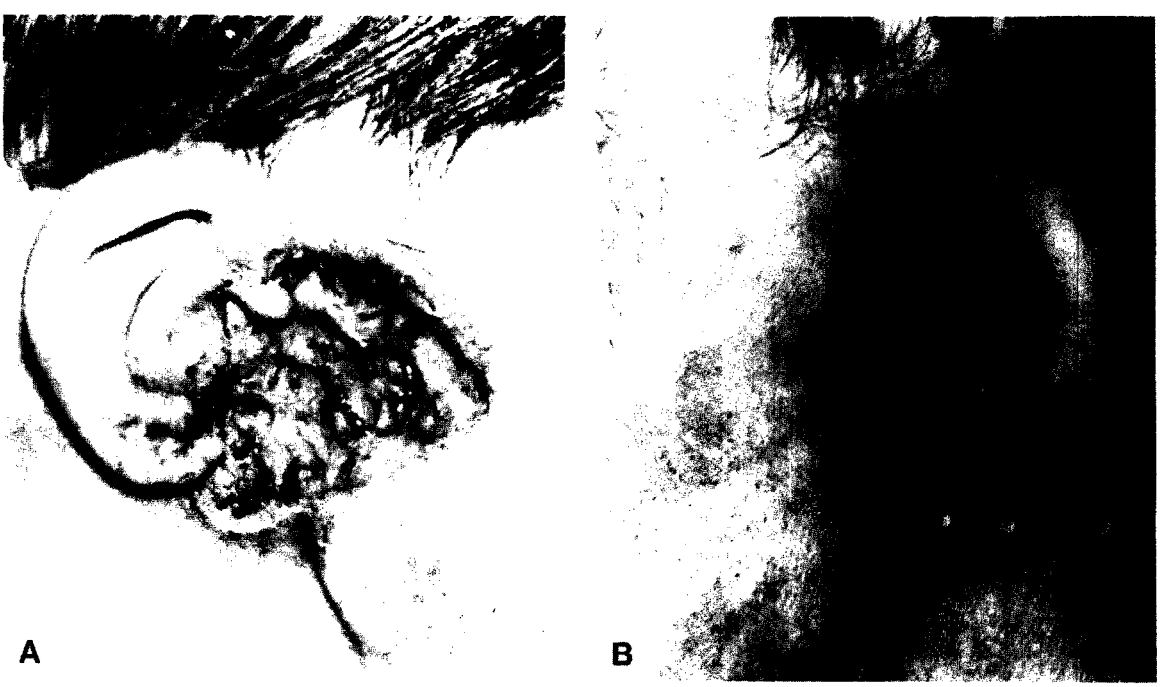

Fig. 2

Figure 2—Extensive basal cell carcinoma of (A) right external auditory meatus (B) left external auditory meatus.

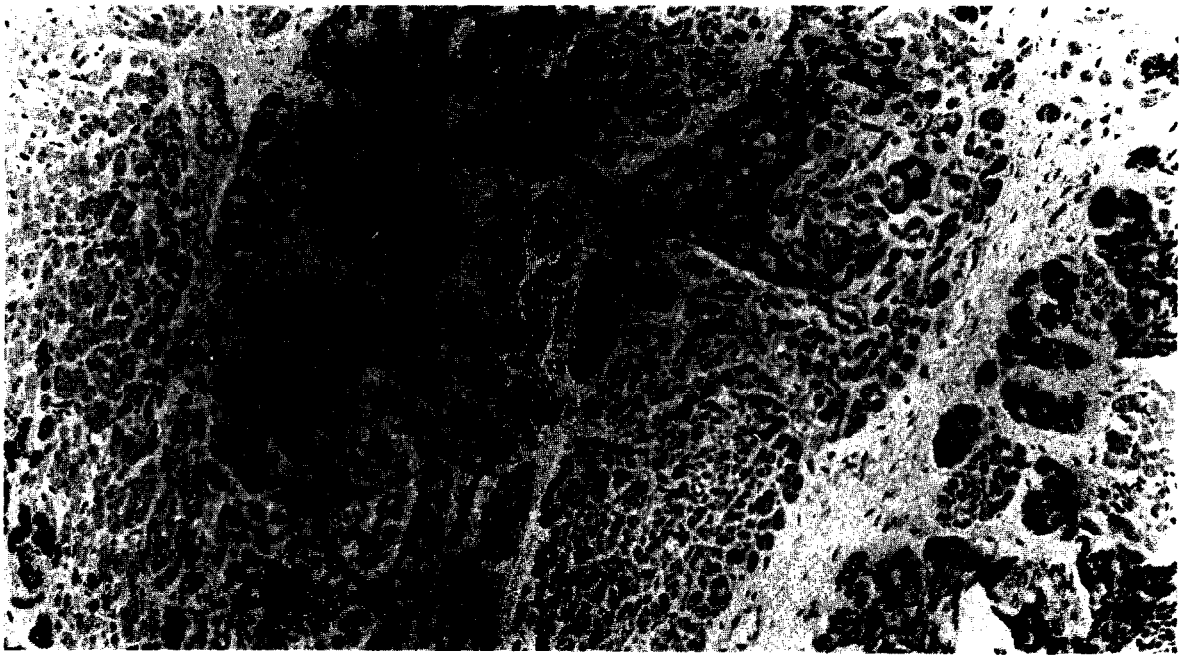

Fig. 3

Fig. 4

Figure 3—Intramyocardial metastases of BCC are present with variable peripheral palisading. (10 ×). Figure 4—Lung metastases showing perivascular and lymphocytic spread of tumour cells with a high degree of peripheral palisading. (4 ×).

2000 rads, the patient himself refused further therapy. Three months later a nodular density (2½ cm) was noted in the left lung field and the possibility of metastasis was considered. The patient was admitted in December 1984 with dehydration and pneumonia which eventually progressed to respiratory failure, sepsis and death. Autopsy revealed extensive BCC of both lungs involving the pleura, the diaphragm, the pericardium and both the epicardium and myocardium (Figs 3 and 4). Metastases were also found at the base of the skull, the ribs and the brain. No sign of other primary malignancy was found.

Discussion

The case described exhibited many of the known features of basal cell naevus syndrome: (i) multiple cutaneous tumours, (ii) jaw cysts, (iii) calcification of the falx cerebri, (iv) ocular hypertelorism, (v) congenital blindness and (vi) skeletal deformity. In addition, we describe diffuse BCC metastases to the pleura, heart and diaphragm. We have also made note of aggressive tumour invasion of bilateral external auditory canals, not uncommon sites for BCC (Goodwin and Jesse, 1980).

Basal cell carcinoma rarely metastasises; from 1894 to 1984 only 175 cases of metastatic BCC were documented in the literature, an incidence of 0.1% (von Domarus and Stevens, 1984). These involved lymph nodes, lungs and bones. A study by Farmer and Helwig (1980) described 17 cases of metastases with the lung as the most commonly affected site (9 cases), followed by bone (5 cases), lymph nodes (4 cases), liver (3 cases), spleen (1 case) and the adrenal gland (1 case). Thirteen of these patients had metastatic lesions involving only one organ system. The mean survival time after metastasis was 1.6 years.

The incidence of metastasis from the basal cell naevus syndrome is not documented; however, metastases have been found at various sites including the lungs (Taylor *et al.*, 1968). To the best of our knowledge, there have been no reports of metastases in this syndrome involving the pleura and diaphragm as well as the pericardium, epicardium and myocardium.

Acknowledgements

The authors gratefully acknowledge the contribution of Minal Patel, MD, Department of Pathology, Mount Sinai Medical Center.

References

Domarus, von H. and Stevens, P. J. (1984). Metastatic basal cell carcinoma. Report of five cases and review of 170 cases in the literature. *Journal of the American Academy of Dermatology*, **10**, 1043.

Farmer, E. R. and Helwig, E. B. (1980). Metastatic basal cell carcinoma: a clinicopathologic study of 17 cases. *Cancer*, **46**, 748.

Fitzpatrick, P. J. and Thompson, G. A. (1982). Gorlin's syndrome or nevoid basal cell carcinoma syndrome. *Journal of the Canadian Medical Assocation*, **127**, 465.

Goodwin, W. J. and Jesse, R. H. (1980). Malignant neoplasms of the external auditory canal and temporal bone. *Archives of Otolaryngology*, **106**, 675.

Gorlin, R. J. and Goltz, R. W. (1960). Multiple nevoid basal cell epithelioma, jaw cysts and bifid rib: a syndrome. *New England Journal of Medicine*, **262**, 908.

Jarisch, W. (1894). Zur Lehre von den Hautgeschewülsten. *Archiv für Dermatologie und Syphilis*, **28**, 163.

Olson, R. A. J., Stroncek, G. G., Scully, J. R. and Govin, L. (1981). Nevoid basal cell carcinoma syndrome: review of the literature and report of a case. *Journal of Oral Surgery*, **39**, 308.

Plaza, de la R., Rodriguez, E. and Castillo, E. (1983). Two cases of nevoid basal cell carcinoma syndrome. *Plastic and Reconstructive Surgery*, **71**, 114.

Satinoff, M. I. and Wells, C. (1969). Multiple basal cell naevus syndrome in ancient Egypt. *Medical History*, **13**, 294.

Taylor, W. B., Anderson, D. E., Howell, J. B. and Thurston, C. S. (1968). The nevoid basal cell carcinoma syndrome: autopsy findings. *Archives of Dermatology*, **98**, 612.

White, J. C. (1894). Multiple benign cystic epitheliomata. *Journal of Cutaneous Diseases*, **12**, 477.

The Authors

Peter A. Winkler, MD, Resident
Bahman Guyuron, MD, Chief
Division of Plastic Surgery, Mount Sinai Medical Center, Cleveland, Ohio.

Requests for reprints to Bahman Guyuron, MD, 29001 Cedar Road: Suite 406, Lyndhurst, Ohio 44124, USA.