Familial malignant melanoma

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Summary—A case is reported of a 28-year-old man with three separate primary cutaneous malignant melanomas. The patient denied any family history of melanoma, but screening of the first degree relatives resulted in the early diagnosis of two malignant melanomas in his brother and the recognition of a strong family history of melanoma.

Figures from many parts of the world have shown a dramatic increase in the incidence of cutaneous malignant melanoma (Elwood and Lee, 1979). In areas such as Queensland with a high incidence (Little et al., 1980), public education about melanoma has led to earlier referral and a consequent improvement in prognosis. By contrast, in Britain there is a low level of public awareness (Doherty et al., 1985), so that treatment is all too frequently delayed.

One group of subjects who have a greatly increased risk of malignant melanoma are relatives of patients with the tumour (Greene and Fraumeni, 1979). Screening of relatives may therefore be of value in the early diagnosis of melanoma.

Case report

A 28-year-old man was referred with a pigmented lesion on the lower abdomen, which had been noted by the GP as an incidental finding on examination for another complaint. The patient had been aware of the lesion for 4 years but had not appreciated its significance. On examination, he was found to have three separate primary malignant melanomas, one on the lower abdomen (Fig. 1), one on the left upper arm (Fig. 2) and one on the left anterior chest; this last lesion also had a number of small satellite tumours around it (Fig. 3). There was marked enlargement of the left axillary lymph nodes. In addition, he had a number of large, atypical, irregular naevi on the trunk and limbs. The melanomas were treated by primary surgical excision, with clearance of the lymph nodes from the left axilla. The diagnosis was confirmed histologically, the lesions on the chest, upper arm and abdomen having Breslow thicknesses of 1.4 mm, 0.61 mm and 1.4 mm respectively. The left axillary
lymph nodes were invaded by metastatic tumour. The naevi were not examined histologically.

The patient denied any family history of melanoma or of atypical or unusual moles, but arrangements were made to review his first degree relatives; his mother, father and one brother were unaffected, but another brother was found to have two primary malignant melanomas, one on the left arm and one on the back, lesions of which he had been hitherto quite unaware. He also had numerous atypical moles on the trunk. The melanomas were excised and the diagnosis of malignant melanoma confirmed histologically; the lesion on the left arm had a Breslow thickness of 1.03 mm and the lesion on the back a Breslow thickness of 0.69 mm. A naevus on the abdomen showed histological features of a dysplastic naevus.

None of the family was aware of any history of malignant melanoma, but further enquiry of other relatives led to the discovery that two maternal uncles had died of the disease in their forties (Fig. 4).

Discussion

Multiple primary lesions in cutaneous malignant melanoma are an uncommon phenomenon. Scheibner et al. (1983) found that of 3128 patients with primary tumours, 90 had more than one primary lesion (2.9%) and only 12 (0.4%) had three or more primary lesions. Moseley et al. (1979) recorded multiple primary lesions in 5.3% of 712 patients with melanoma, three or more lesions being present in 14 (1.9%). By contrast, multiple primary lesions are very much more common when there is a family history of malignant melanoma. Wallace et al. (1971) found multiple primary lesions in 8 of 58 patients with a family history (14%) compared to only 49 multiple primaries in 1458 patients without a family history (3.4%).

The phenomenon of familial clustering of malignant melanoma has long been recognised and overall it has been estimated that there is a family history of the tumour in 11% or more of all cases (Wallace et al., 1971; Greene and Fraumeni, 1979). Our understanding of familial malignant melanoma was clarified by the work of Reimcr et al. (1978) who described seven families, in each of which more than one member was affected by it. They observed that of 20 patients with melanoma, 18 also had numerous naevi of a clinically and histologically distinctive form. In addition, of unaffected first degree relatives of melanoma patients in these families, 56% had these atypical or “dysplastic” naevi. The authors termed the disorder the Dysplastic Naevus Syndrome and showed that it appeared to be inherited as an autosomal dominant trait. In a subsequent prospective study of 14 affected pedigrees, 39 new cases of malignant melanoma were found in an 8-year period, all occurring in patients with dysplastic naevi. Furthermore, in patients with the dysplastic naevus syndrome, multiple primary lesions are not uncommon. Greene et al. (1985) found multiple primary lesions in 21 of 69 patients (30%) compared to only 3 of 100 sporadic cases of malignant melanoma. Such a finding is in accord with earlier observations of an increased incidence of multiple primary lesions in familial cases of melanoma.

Clearly, therefore, relatives of patients with melanoma arising in the context of the dysplastic naevus syndrome merit screening for melanoma. It

![Figure 4 — Family tree.](Fig. 4)
is now evident, however, that this syndrome shows a greater degree of phenotypic variation than was initially appreciated. Obligate carriers (i.e. those with both a parent and a child affected) may be unaffected, and patients with malignant melanoma and a family history of dysplastic naevus syndrome may themselves lack dysplastic naevi (Lynch et al., 1983).

Family histories are often incomplete or inaccurate. Given the prognostic significance of early diagnosis in malignant melanoma, we believe that screening of families of patients with this tumour is of value and may bring a significant yield of early cases. Although family screening is especially important where there is a history of multiple primary lesions or evidence of dysplastic naevi, we believe that, given the marked phenotypic variation within the spectrum of the dysplastic naevus syndrome, the screening of all first degree relatives of patients with cutaneous malignant melanoma merits consideration.

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References


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