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## Summary and Introduction

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### Summary

**Background:** Desmoplastic melanoma (DM) is an uncommonly encountered type of malignant melanoma. The clinical appearance of DM can be highly variable and thus, diagnosis of this tumour is difficult and very often may mislead the physician.

**Objectives:** To make a critical review of the contemporary literature on DM, to pool the data from published studies and to evaluate the clinical and morphological characteristics of this neoplasm.

**Methods:** All studies or reports on DM including 10 or more participants with reported clinical and histological characteristics of the tumour were included.

**Results:** In the 17 studies that met the inclusion criteria a total of 856 patients with DM was reported. There was a male predilection, with a male/female ratio of almost 2 : 1 (63% of the lesions were diagnosed in males). The head and neck were the most common sites of DM for both sexes (53.2%). The data confirmed that DM usually has an advanced Breslow thickness at the time of presentation. Histopathological diagnosis of DM is sometimes difficult and the absence of pigmentation is probably the major cause for failure to recognize DM histologically. The pooled data from included studies showed that the incidence of nodal metastasis is lower in patients with DM than in patients with other forms of cutaneous melanoma.

**Conclusions:** Prompt definitive surgical excision is the treatment of choice for DM. Improved knowledge of the clinical behaviour and histological features of DM is important for more effective management of patients with DM.

### Introduction

Malignant melanoma may express a variety of phenotypes with different clinical and cytomorphological features. Desmoplastic melanoma (DM) is an uncommonly encountered type of malignant melanoma, first described by Conley *et al.* in 1971 as an invasive melanoma composed of spindle cells surrounded by abundant collagen.<sup>[1]</sup> Subsequently, Reed and Leonard extended this definition to include neurotropic malignant melanoma as a variant of DM that showed neurotropism or underwent neural differentiation.<sup>[2]</sup> Nowadays DM is usually said to comprise two subtypes: DM with neurotropism and DM without neurotropism.<sup>[3]</sup>

The clinical appearance of DM can be highly variable and DM may mimic a variety of different lesions. Usually it has an innocuous clinical appearance and it is described as an indurated discoid papule, plaque or nodule. Pigmentation is frequently absent, although a lentigo or lentigo maligna-like discoloration adjacent to the nodule is not uncommon.<sup>[4]</sup> Hence, the clinical impression at presentation may vary from that of basal cell or squamous cell carcinomas, dermatofibromas or sarcomas to cysts and indurated plaque-like lesions resembling scars.<sup>[5]</sup> DM has high propensity for local recurrence and although lesions may show only minor cytological atypia, DM in fact may have an aggressive behaviour. Clinical diagnosis of this tumour is difficult and very often may mislead the physician.

DM does not show the conventional microscopic features of melanoma. Microscopically, DMs present as large, poorly circumscribed neoplasms of variable size, that in some cases extend into subcutaneous tissue, fascia and nerves.<sup>[5]</sup> DM is characterized histologically by dermal and/or subcutaneous infiltrates of spindle-shaped cells arranged singly or in thin fascicles within a prominent collagenous or, less commonly, myxoid stroma. Thus, DM can be difficult to diagnose microscopically by conventional haematoxylin and eosin staining. Immunohistochemical staining for S100 protein is often recommended as a valuable adjunct in the diagnosis of DM.<sup>[6]</sup>

Due to the rarity of DM, extensive epidemiological evaluation has not been possible and only studies with small numbers of patients have been published. We have extracted and pooled data from published studies on DM in order to understand better the clinical and morphological characteristics of this neoplasm.

### Materials and Methods

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The aim was to identify all relevant papers on DM available by January 2004. All studies that met the following criteria were included: clinical studies or case series reports of patients diagnosed with DM; studies or reports including 10 or more participants and reporting clinical and histological characteristics of the tumour.

A computerized search was performed to identify all registered articles on the subject published by January 2004. Studies

were identified through sensitive electronic searches of Medline (from 1966 to January 2004) and Embase (from 1974 to January 2004) using the recommended Cochrane Collaboration search strategy with MeSH headings 'melanoma' and 'desmoplastic' including all subheadings. The reference lists of all relevant retrieved papers and review articles were checked to identify other eligible studies not found in computerized database searching. No language restriction was applied. Unpublished studies were not included.

The following data were extracted independently from each included study: basic characteristics (total number of patients, number of patients according to sex, mean age of patients and age range, median follow-up and range), body location of the DM, basic histological characteristics (Breslow thickness, Clark level, presence of neurotropism and amelanosis) and data on tumour recurrences and survival (local, regional and systemic metastases, overall survival and disease-free survival).

## Results

We retrieved 222 published papers reporting DM. Seventeen papers met our inclusion criteria and are included in our pooled analysis.<sup>[7-23]</sup> Basic characteristics of the included studies are presented in Table 1. The number of patients enrolled in the studies ranged from 11 to 280. In total, 856 patients with a diagnosis of DM were evaluated in the 17 included studies. There were 539 males (63.0%) and 317 females (37.0%). Mean age of patients varied from 55.6 to 71.3 years across the included studies while the age range was 4-99 years.

**Table 1. Basic Characteristics of the Included Studies**

Study	Year	Number of patients			Age of patients (years)		Follow-up (months)	
		Total	Male n (%)	Female n (%)	Mean	Range	Median	Range
Gyorki <i>et al.</i> <sup>[7]</sup>	2003	27	20 (74.1)	7 (25.9)	64 <sup>a</sup>	35-83	27	9-64
Vongtama <i>et al.</i> <sup>[8]</sup>	2003	44	33 (75.0)	11 (25.0)	66	35-87	45	11-145
Payne <i>et al.</i> <sup>[9]</sup>	2001	30	21 (70.0)	9 (30.0)	63	33-87	18 <sup>b</sup>	2-57
Jaroszewski <i>et al.</i> <sup>[10]</sup>	2001	59	37 (62.7)	22 (37.3)	62.8	31-91	46 <sup>b</sup>	1-195
Thelmo <i>et al.</i> <sup>[11]</sup>	2001	16	11 (68.8)	5 (31.2)	57.5	34-84	3	1-43
Wharton <i>et al.</i> <sup>[12]</sup>	1999	18	8 (44.4)	10 (55.6)	64.1	30-87	17	6-114
Quinn <i>et al.</i> <sup>[13]</sup>	1998	280	178 (63.6)	102 (36.4)	59.6	24-91	NR	NR
Rutten <i>et al.</i> <sup>[14]</sup>	1996	34	12 (35.3)	22 (64.7)	71.3	50-92	36	24-84
Carlson <i>et al.</i> <sup>[15]</sup>	1995	28	15 (53.6)	13 (46.4)	59	22-83	24	5-132
Skelton <i>et al.</i> <sup>[16]</sup>	1995	126	84 (66.7)	42 (33.3)	63	4-99	NR	NR
Weinzweig <i>et al.</i> <sup>[17]</sup>	1995	11	7 (63.6)	4 (36.4)	68.4	48-81	57.9 <sup>b</sup>	1-161
Anstey <i>et al.</i> <sup>[18]</sup>	1993	25	14 (56.0)	11 (44.0)	65	39-86	47 <sup>b</sup>	9-120
Smithers <i>et al.</i> <sup>[19]</sup>	1992	58	33 (56.9)	25 (43.1)	63 <sup>a</sup>	17-89	30	1-124
Beenken <i>et al.</i> <sup>[20]</sup>	1989	16	14 (87.5)	2 (12.5)	62.7	42-76	36	3-140
Jain and Allen <sup>[21]</sup>	1989	45	31 (68.9)	14 (31.1)	64	42-91	55.2 <sup>b</sup>	4-180
Egbert <i>et al.</i> <sup>[22]</sup>	1988	25	11 (44.0)	14 (56.0)	61.2	38-83	32.4 <sup>b</sup>	1.2-108
Walsh <i>et al.</i> <sup>[23]</sup>	1988	14	10 (71.4)	4 (28.6)	55.6	17-77	55	5-276

NR, not reported.

<sup>a</sup>Median.

<sup>b</sup>Mean.

Data on the location of DM were available for 836 patients (97.7%) ( Table 2 ). The lesions were most commonly located

on the head and neck (53.2%), followed by the extremities (26.2%) and the trunk (20.6%). Three studies reported the total number of patients with limb DM, but did not report number of patients with DM separately for the lower and upper limbs. Of 188 patients with limb DM for whom the data were available, 132 (70.2%) had DM located on the upper limbs, while 56 (29.8%) had lower-limb DM.

**Table 2. Location of Desmoplastic Melanoma**

Study	Head and neck		Trunk		Extremities					
	n	%	n	%	Total		Upper		Lower	
					n	%	n	%	n	%
Gyorki <i>et al.</i> <sup>[7]</sup>	14	51.9	8	29.6	5	18.5	NR	NR	NR	NR
Vongtama <i>et al.</i> <sup>[8]</sup>	30	68.2	6	13.6	8	18.2	5	11.4	3	6.8
Payne <i>et al.</i> <sup>[9]</sup>	12	40.0	10	33.3	8	26.7	5	16.7	3	10.0
Jaroszewski <i>et al.</i> <sup>[10]</sup>	36	61.0	11	18.6	12	20.4	NR	NR	NR	NR
Thelmo <i>et al.</i> <sup>[11]</sup>	7	43.8	4	25.0	5	31.2	4	25.0	1	6.2
Wharton <i>et al.</i> <sup>[12]</sup>	12	66.6	3	16.7	3	16.7	3	16.7	0	0
Quinn <i>et al.</i> <sup>[13]</sup>	106	38.9	67	24.5	101	36.6	69	25.2	32	11.4
Rutten <i>et al.</i> <sup>[14]</sup>	27	81.8	5	15.2	1	3.0	1	3.0	0	0
Carlson <i>et al.</i> <sup>[15]</sup>	21	75.0	5	17.9	2	7.1	2	7.1	0	0
Skelton <i>et al.</i> <sup>[16]</sup>	61	50.4	21	17.4	39	32.2	30	24.8	9	7.4
Weinzweig <i>et al.</i> <sup>[17]</sup>	4	36.4	4	36.4	3	27.2	1	9.1	2	18.1
Anstey <i>et al.</i> <sup>[18]</sup>	18	72.0	3	12.0	4	16.0	1	4.0	3	12.0
Smithers <i>et al.</i> <sup>[19]</sup>	24	41.4	20	34.5	14	24.1	NR	NR	NR	NR
Beenken <i>et al.</i> <sup>[20]</sup>	13	81.3	2	12.5	1	6.2	1	6.2	0	0
Jain and Allen <sup>[21]</sup>	35	77.8	5	11.1	5	11.1	3	6.7	2	4.4
Egbert <i>et al.</i> <sup>[22]</sup>	21	84.0	1	4.0	3	12.0	3	12.0	0	0
Walsh <i>et al.</i> <sup>[23]</sup>	4	28.6	5	35.7	5	35.7	4	28.6	1	7.1
Total	445	53.2	172	20.6	219	26.2				

NR, not reported.

Histological characteristics were not completely reported in all included studies ( Table 3 ). Twelve studies reported data on the Breslow thickness of the primary DM. The mean Breslow thickness of the original DM ranged from 2.0 to 6.5 mm. The lowest Breslow thickness reported among patients from included studies was 0.2 mm, while the highest Breslow thickness was 18 mm. Clark level was fully reported only in eight studies. Most tumours were Clark level IV or V.

**Table 3. Histological Characteristics of Desmoplastic Melanoma**

Study	Mean (range) Breslow thickness (mm)	Clark level (%)			Neurotropism (%)	Amelanosis (%)
		≤ III	IV	V		
Gyorki <i>et al.</i> <sup>[7]</sup>	2.2 <sup>a</sup> (0.7-20)	0	74.1	25.9	37.0	-
Vongtama <i>et al.</i> <sup>[8]</sup>	-	20.5	54.5	25.0	47.7	-
Payne <i>et al.</i> <sup>[9]</sup>	2.6 (0.5-9.5)	-	-	-	16.7	-

Jaroszewski <i>et al.</i> <sup>[10]</sup>	6.5 (0.5-30)	-	-	-	0	-
Thelmo <i>et al.</i> <sup>[11]</sup>	3.9 (1.4-11.0)	0	68.8	31.2	43.8	-
Wharton <i>et al.</i> <sup>[12]</sup>	2.0 (0.8-3.9)	11.1	55.6	33.3	77.8	50.0
Quinn <i>et al.</i> <sup>[13]</sup>	2.5 <sup>a</sup> (0.2-18)	10.8	55.4	33.8	32.1	85.0
Rutten <i>et al.</i> <sup>[14]</sup>	3.8 (1.0-11.0)	0	55.9	29.4 14.7 <sup>b</sup>	50.0	60.0
Carlson <i>et al.</i> <sup>[15]</sup>	4.8 (0.3-9.0)	3.8	48.1	48.1	46.4	57.1
Weinzweig <i>et al.</i> <sup>[17]</sup>	-	-	-	-	-	72.7
Anstey <i>et al.</i> <sup>[18]</sup>	4.9 (1.0-10.1)	-	-	-	36.0	80.0
Smithers <i>et al.</i> <sup>[19]</sup>	4.3 <sup>a</sup> (0.45-16)	-	-	-	41.4	70.7
Beenken <i>et al.</i> <sup>[20]</sup>	5.1 (1.4-13.0)	-	-	-	-	-
Jain and Allen <sup>[21]</sup>	2.5 (0.5-6.0)	-	-	-	26.7	93.3
Egbert <i>et al.</i> <sup>[22]</sup>	-	-	-	-	24.0	92.0
Walsh <i>et al.</i> <sup>[23]</sup>	-	7.7	69.2	23.1	71.4	46.2

<sup>a</sup>median.

<sup>b</sup>Clark IV/V. -, not reported.

The presence of neurotropism was reported on in 14 studies. One study excluded patients with DM with neurotropism.<sup>[10]</sup> The percentage of DMs with neurotopism ranged from 16.7% to 77.8% among the included studies. Ten studies reported the absence of pigmentation in the primary lesion. The percentage of DMs with amelanosis ranged from 46.2% to 93.3% among the included studies.

Data on follow-up were available for 15 studies. Follow-up ranged from 1 to 276 months. Six studies reported mean follow-up (range 18-57.9 months) while nine studies reported median follow-up (range 3-55 months).

Data on recurrent disease were available for a total of 703 patients ( Table 4 ). One study did not report the data on recurrences.<sup>[16]</sup> In only one study was no recurrent disease detected at all,<sup>[11]</sup> although the follow-up of the patients in this study was very short (median 3 months). Only one study reported an incidence of systemic disease higher than the incidence of local recurrence.<sup>[7]</sup> In the 14 remaining studies the most common type of recurrence was local, the rate ranging from 6.7% to 56.0%.

**Table 4. Sites of Recurrence for Desmoplastic Melanoma and Survival Data**

Study	n <sup>a</sup>	Local n (%)	Regional n (%)	Systemic n (%)	Death	OS (%)	DFS (%)
Gyorki <i>et al.</i> <sup>[7]</sup>	27	1 (3.7)	0	4 (14.8)	2 (7.4)	92.2	85.2
Vongtama <i>et al.</i> <sup>[8]</sup>	44	21 (47.7)	3 (6.8)	17 (38.6)	5 (11.4)	88.6	79.5
Payne <i>et al.</i> <sup>[9]</sup>	27	7 (25.9)	3 (11.1)	2 (7.4)	1 (3.7)	96	NR
Jaroszewski <i>et al.</i> <sup>[10]</sup>	59	23 (39.0)	1 (1.7)	16 (27.1)	13 (22.0)	88.0	72.9
Thelmo <i>et al.</i> <sup>[11]</sup>	16	0	0	0	0	100	100
Wharton <i>et al.</i> <sup>[12]</sup>	15	1 (6.7%)	0	0	1 (6.7)	93.3	93.3
Quinn <i>et al.</i> <sup>[13]</sup>	280	38 (13.6)	26 (9.3)	39 (13.9)	NR	75.2 <sup>b</sup>	62 <sup>b</sup>
Rutten <i>et al.</i> <sup>[14]</sup>	22	7 (31.8)	NR	4 (18.2)	5 (22.7)	77.3	NR
Carlson <i>et al.</i> <sup>[15]</sup>	26	7 (26.9)	4 (15.4)	3 (11.5)	3 (11.5)	88.5	80.8

Weinzweig <i>et al.</i> <sup>[17]</sup>	11	6 (54.5)	1 (9.1)	3 (27.2)	3 (27.2)	72.8	63.6
Anstey <i>et al.</i> <sup>[18]</sup>	25	14 (56.0)	NR	8 (32.0)	11 (44.0)	56.0	52.0
Smithers <i>et al.</i> <sup>[19]</sup>	58	17 (29.3)	8 (13.8)	12 (20.7)	12 (20.7)	79.3	74.1
Beenken <i>et al.</i> <sup>[20]</sup>	16	8 (50.0)	3 (18.8)	7 (43.8)	4 (25.0)	75.0	62.5
Jain and Allen <sup>[21]</sup>	42	23 (54.8)	NR	17 (40.5)	18 (42.9)	67.1	57.1
Egbert <i>et al.</i> <sup>[22]</sup>	23	12 (52.2)	1 (4.3)	5 (21.7)	5 (21.7)	78.3	73.9
Walsh <i>et al.</i> <sup>[23]</sup>	12	6 (50.0)	NR	2 (16.7)	2 (16.7)	83.3	83.3

OS, overall survival at the end of follow-up; DFS, disease-free survival at the end of follow-up; NR, not reported.

<sup>a</sup> *n*, number of patients for whom follow-up details were available.

<sup>b</sup> Five-year survival.

In contrast to the pattern seen generally for melanoma, the development of regional metastases was rare, incidence ranging from 0% to 18.8%. Systemic metastatic disease occurred in patients from 14 studies. The incidence of systemic metastases ranged from 7.4% to 43.8%. In the pooled analysis of 703 patients, 191 (27.2%) suffered from local tumour recurrences. Regional lymph node metastases occurred in 50 patients (7.1%), while systemic metastases developed in 139 patients (19.8%).

One study<sup>[13]</sup> reported 5-year overall survival (OS) and disease-free survival (DFS) (75.2% and 62%, respectively). Sixteen studies reported OS at the end of the follow-up period or had data from which we were able to calculate OS at the end of follow-up. This ranged from 67.1% to 100%. Fourteen studies reported DFS at the end of the follow-up period or had data from which we were able to calculate DFS at the end of follow-up. This ranged from 52% to 100%. Data on OS and DFS are presented in Table 4.

**Table 4. Sites of Recurrence for Desmoplastic Melanoma and Survival Data**

Study	<i>n</i> <sup>a</sup>	Local <i>n</i> (%)	Regional <i>n</i> (%)	Systemic <i>n</i> (%)	Death	OS (%)	DFS (%)
Gyorki <i>et al.</i> <sup>[7]</sup>	27	1 (3.7)	0	4 (14.8)	2 (7.4)	92.2	85.2
Vongtama <i>et al.</i> <sup>[8]</sup>	44	21 (47.7)	3 (6.8)	17 (38.6)	5 (11.4)	88.6	79.5
Payne <i>et al.</i> <sup>[9]</sup>	27	7 (25.9)	3 (11.1)	2 (7.4)	1 (3.7)	96	NR
Jaroszewski <i>et al.</i> <sup>[10]</sup>	59	23 (39.0)	1 (1.7)	16 (27.1)	13 (22.0)	88.0	72.9
Thelmo <i>et al.</i> <sup>[11]</sup>	16	0	0	0	0	100	100
Wharton <i>et al.</i> <sup>[12]</sup>	15	1 (6.7%)	0	0	1 (6.7)	93.3	93.3
Quinn <i>et al.</i> <sup>[13]</sup>	280	38 (13.6)	26 (9.3)	39 (13.9)	NR	75.2 <sup>b</sup>	62 <sup>b</sup>
Rutten <i>et al.</i> <sup>[14]</sup>	22	7 (31.8)	NR	4 (18.2)	5 (22.7)	77.3	NR
Carlson <i>et al.</i> <sup>[15]</sup>	26	7 (26.9)	4 (15.4)	3 (11.5)	3 (11.5)	88.5	80.8
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Anstey <i>et al.</i> <sup>[18]</sup>	25	14 (56.0)	NR	8 (32.0)	11 (44.0)	56.0	52.0
Smithers <i>et al.</i> <sup>[19]</sup>	58	17 (29.3)	8 (13.8)	12 (20.7)	12 (20.7)	79.3	74.1
Beenken <i>et al.</i> <sup>[20]</sup>	16	8 (50.0)	3 (18.8)	7 (43.8)	4 (25.0)	75.0	62.5
Jain and Allen <sup>[21]</sup>	42	23 (54.8)	NR	17 (40.5)	18 (42.9)	67.1	57.1
Egbert <i>et al.</i> <sup>[22]</sup>	23	12 (52.2)	1 (4.3)	5 (21.7)	5 (21.7)	78.3	73.9
Walsh <i>et al.</i> <sup>[23]</sup>	12	6 (50.0)	NR	2 (16.7)	2 (16.7)	83.3	83.3

OS, overall survival at the end of follow-up; DFS, disease-free survival at the end of follow-up; NR, not reported.

<sup>a</sup> *n*, number of patients for whom follow-up details were available.

<sup>b</sup> Five-year survival.

## Discussion

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DM is an uncommon form of melanoma. Thus, it is important to understand its clinical characteristics and morphological patterns better as the experience of it in any one institution is low. There is a lack of statistically significant studies on the behaviour of DM due to the small number of patients included in the studies, patients' advanced age and short follow-up. Our study pooled the data from all published studies evaluating the characteristics of DM and thus represents the currently largest sample size.

DM is easily missed or misdiagnosed in the early stage, as it is often clinically innocuous and quite distinct from other types of melanoma.<sup>[4,24]</sup> We confirm in this study that DM usually has an advanced Breslow thickness at the time of presentation, which probably results from the difficulties in clinical diagnosis.

This analysis shows that patients diagnosed with DM are older (mean age around 63 years) than patients with unselected melanoma (the mean age for patients with superficial spreading melanoma is mid-40s). There is a male predilection, with a male/female ratio of almost 2 : 1.

The head and neck were the most common sites of DM for both sexes, although it can be also located on the trunk and upper limbs and in less expected sites such as the lower limbs, oral or genital mucosa. An increased association with lentigo maligna melanoma may support the possible link of DM with sun exposure.<sup>[25]</sup>

Histopathological diagnosis of DM is sometimes difficult and represents a significant challenge to the pathologist. The absence of pigmentation is probably the major cause of failure to recognize DM as melanoma histologically. Careful pathological examination with a critical review of multiple sections is required in order to avoid failure in the diagnosis of DM.<sup>[6,26]</sup> The use of immunohistochemistry (testing for S100 antigen) is suggested as a useful tool in establishing the diagnosis.

Close follow-up is necessary to detect recurrences and metastatic disease in all patients with melanoma including those with DM. Follow-up of patients with DM showed that DM is characterized by a high incidence of local recurrence, low incidence of lymph node metastases, and a propensity to develop systemic metastases. This is in contrast to other types of cutaneous melanoma where local recurrence is rare and the probability of lymph node metastases is mainly related to the thickness of the lesion.

Review of the published studies showed that local recurrence in DM is associated with an increased risk of developing additional local recurrences and an increased risk of developing systemic metastases. The high incidence of local recurrence after excision of the primary DM may be attributed to the clinical misdiagnosis of lesions, difficulty in the pathological examination and interpretation of specimens, inadequate surgical excision, high Breslow thickness of the tumour at time of diagnosis, and the neurotropic nature of infiltration.<sup>[10]</sup>

Our pooled analysis showed that the incidence of local recurrence among studies in which the mean Breslow thickness was < 4.0 mm was 19.5%, while among studies with the mean Breslow thickness > 4.0 mm the rate of local recurrence was 40.2%.

It was suggested that neurotropism is related to an increase in the frequency of local recurrences.<sup>[27]</sup> Also, it seems that neurotropism is associated with a significant decrease in survival in patients with DM.<sup>[27]</sup> However, we were unable to establish the association between neurotropism and local recurrence rate and survival due to the lack of the relevant raw data from the studies included in our analysis.

Although DMs are thicker lesions at the time of presentation compared with other variants of melanoma, the incidence of nodal metastasis is lower than in other forms of cutaneous melanoma where the incidence of regional metastasis as the site of first clinical relapse ranges from approximately 45% to 65% of all recurrences and correlates with the Breslow thickness of the primary tumour.<sup>[28]</sup> Our study demonstrated that lymph node involvement was present in 7.1% of the patients enrolled in included studies, showing no correlation with the Breslow thickness of the primary DM. The low

incidence of lymph node metastasis in line with that seen in patients with soft tissue sarcoma suggests a different biology and natural history for DM compared with other types of melanoma. Indeed, DMs behave in a similar way to malignant schwannoma, from which it may also be difficult to make a histological distinction.

Systemic metastases appear to be related to previous recurrences and to depth of lesion. The lung is the most common site of systemic metastases. Some data suggest that DM may have an improved prognosis over the other forms of cutaneous melanoma. In other studies the survival from DM was not significantly different than from conventional melanoma. The aggressive clinical behaviour of DM seen in these studies can be attributed to the misdiagnosis of the early tumours and inadequate assessment of margins.

Prompt surgical excision of DM is the treatment of choice as it may decrease the chance of multiple local recurrences requiring multiple resections, or even evolution into metastatic disease. Although currently there is a trend towards narrower excision margins for cutaneous melanoma, optimal margins for DM are not established. Because of the unique microscopic and clinical behaviour of DM, routine recommendations for surgical resection of the more common melanomas may not apply to DM.<sup>[24]</sup> Many surgeons advise a minimum clearance margin of at least 1 cm. The problem with DM is that most DMs are invasive to a significant degree by the time of diagnosis, which makes it difficult to achieve clear surgical margins on an initial resection, especially if the tumour is present in an area in which large excisions are difficult functionally and cosmetically, such as on the face.

The low incidence of regional lymph node metastases in patients with DM suggests that elective lymph node dissection is not indicated in these patients. Recent studies advocate the use of sentinel lymph node biopsy (SLNB) and selective lymph node dissection as the most appropriate method for evaluation of the lymph node basins.<sup>[10]</sup> However, there is insufficient evidence on this matter and a multicentre study is needed to assess the clinical significance of SLNB in patients with DM.

The use of radiation therapy, adjunctive chemotherapy and immunotherapy has not been sufficiently studied in patients with DM. Some authors recommended adjuvant postoperative radiation therapy as a part of treatment of DM, claiming that it may be beneficial for local control as it can reduce the high rate of local recurrence of DM after surgical resection.<sup>[8,29]</sup> However, the number of patients with DM treated with one of these modalities is too small to establish a strong clinical judgement when considering postsurgical treatment of a patient with DM.

This review has highlighted the distinctive clinical behaviour and histological features of DM. Better knowledge of this type of melanoma should lead to its improved and more effective management.

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