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Melanosis Naeviformis of Becker and Scoliosis: A Coincidence?

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Melanosis naeviformis of Becker (MNB) can be associated with hypoplasia of soft tissue or extremities, spina bifida and scoliosis of the vertebral column. We have investigated 50 patients (42 men, 8 women) with MNB radiologically. Scoliosis was diagnosed in 13 patients (26%). The curves of scoliosis varied from 11° to 17°. Physical examination revealed no gross asymmetries of the trunk, extremities or breasts. No correlation was seen between the age of patients and the scoliotic curve. In one family the father and oldest son had MNB with scoliosis and the other son and daughter had MNB without scoliosis.

Since only mild scoliosis is found in patients with MNB, X-ray examination of the vertebral column has no therapeutic consequences. ***Key words: familial melanosis naeviformis of Becker; X-ray.***

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Melanosis naeviformis of Becker (MNB) is known as an unilateral hyperpigmented macula with hypertrichosis and is most characteristically situated on the shoulder, anterior chest, scapular region or arm (1). It mainly appears in puberty or late adolescence and occurs more frequently in males than in females (M:F ratio=6:1) (2). An incidence of 0.5–3% has been reported (2, 3). MNB occurs sporadically, although familial occurrence has been reported in different families (4–6). According to Happle, MNB is an example of a phenotype with paradominant inheritance (7). This means that the trait only becomes manifest when a somatic mutation occurs in the developing embryo, which might also explain why MNB is associated with different abnormalities. Commonly associated abnormalities include hypoplasia of soft tissue or extremities, spina bifida and scoliosis of the vertebral column. Glinick et al. (2) suggest that MNB might be a marker for underlying structural anomalies. It is remarkable that these abnormalities are observed more frequently in female patients (M:F ratio= 7:9). Different associated abnormalities have been studied; however no investigation has been made of a possible correlation between MNB and scoliosis.

In order to evaluate MNB and the incidence of soft tissue or bony abnormalities, especially scoliosis, we studied 50 patients with MNB.

PATIENTS AND METHODS

During a period of 2 years, 50 patients (6–61 years, average age 26.1 years) with MNB who visited the outpatient clinic of the academic hospitals in Maastricht (AZM) and Amsterdam (AMC) were selected for the study. All patients with a clinically characteristic MNB were included. In all patients the naevus was located either on the trunk and/or shoulder. All patients were physically examined for soft tissue abnormalities. From all patients an antero-posterior and lateral radiograph of the vertebral column was obtained, and radiological examination was made to estimate the presence of scoliosis. This was done according to the method developed by Cobb (8).

In Cobb's method the end vertebral bodies with the greatest tilt towards the horizontal are determined on a spinal radiograph (9). The lines perpendicular to the endplates of these vertebrae are used to construct the scoliosis angle. The measurements of these curves were performed by two independent radiologists. A lateral curve with an angle of 10° or more is defined as scoliosis by the Scoliosis Research Society (10).

RESULTS

Scoliosis was diagnosed in 11 out of 42 men (26.2%), and 2 out of 8 women (25%). The sex ratio of the scoliosis group was 5.5:1, M:F. Curves of scoliosis varied from 11° to 17° (Table I). Physical examination revealed no gross asymmetries of the trunk or hypoplasia of breasts or hyperplasia of extremities. The age of patients with MNB and scoliosis (29.7 years) was not significantly higher in the group without scoliosis (25.1 years) ($p=0.07$). No correlation existed between the age of patients and scoliotic curve. In the group of patients that were 30 years or older (16 patients), scoliosis was diagnosed in 6 cases.

A variety of congenital anomalies of the vertebral column and ribs were observed. These consisted of lumbalisation of S_1 (7 patients), spina bifida occulta of L_5 or S_1 (6 patients), sacralisation of L_5 (2 patients), accessory pair of ribs (2 patients), spondylolysis (1 patient), hemisacralisation of L_5 (2 patients), rudimentary rib of Th_{12} or L_1 (2 patients).

In one family the father and oldest son had MNB with scoliosis, and a son and daughter had MNB without scoliosis. In another family a sister and brother both had MNB but without scoliosis.

Table I. *Vertebral curves (in degrees) of patients with scoliosis and MNB, their age in years and sex*

Age	Scoliotic curve (°)	Sex
19	12	F
20	12	M
20	16	M
21	15	M
23	11	M
23	11	M
25	11	M
30	12	M
34	17	M
35	14	M
43	17	F
46	13	M
47	15	M

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DISCUSSION

MNB is associated with different abnormalities, including hypoplasia of the ipsilateral and contralateral breast, ipsilateral arm shortening, ipsilateral foot enlargement, scoliosis, lumbar spina bifida, pectus carinatum and accessory scrotum (2, 11–14). Most abnormalities are described in case reports.

Two studies of larger groups of patients with MNB were made by Miura & Takemura and Tymen et al., to investigate sensibility and clinical features, respectively (15, 16). In most patients with MNB a diminished sensibility in the naevus was found (15). The clinical expression of MNB varies more than the original description (16). As far as we know no studies have investigated the incidence of scoliosis and MNB.

Scoliosis is defined as a deformity of the trunk, characterized by a lateral curve in the vertebral column of 10° or more (17). Idiopathic scoliosis is seen much more in females.

In young patients therapy seems useful to prevent progress of scoliosis (9). Some authors recommend treating idiopathic scoliosis in young patients with a curve $>20°$ with a brace (3).

Our results show a strong correlation between MNB and scoliosis: 26% of patients had a curve $>10°$, in contrast to 2% in the normal population (9, 17). Remarkable is the equal sex distribution of the scoliosis in patients with MNB, in contrast to the female predominance in idiopathic scoliosis; sex ratio F:M=9:1.

Although no correlation between age and scoliosis can be established, there seems to be a tendency that the scoliotic curve increases with age ($r=0.19$, $p=0.06$). This might be explained by the progression of the curves after skeletal maturity due to aging (9).

Familial occurrence of MNB has been reported. It has never been reported, however, that four members of one family have MNB. Father and son have scoliosis as well as MNB. Familial occurrence of both MNB and scoliosis supports the theory of Happle that MNB can be explained by paradominant inheritance.

Our results show a strong correlation between MNB and scoliosis. Because (early) diagnosis of mild scoliosis has no therapeutic consequences and does not influence the prognosis, the indication to examine the vertebral column of all patients with MNB radiologically is doubtful. However, it is advisable to examine the patient clinically for a lateral gibbus in order to select risk patients for scoliosis.

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