BATHING TRUNK NEVUS ASSOCIATED WITH NEUROFIBROMATOSIS AND RAISED INTRACRANIAL TENSION

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A 17-year-old boy had bathing trunk nevus with multiple large neurofibromata on the nevus and raised intracranial tension presenting with bilateral papilloedema and pyramidal tract signs. This combination of features is extremely rare.

Key words : Bathing trunk nevus, Neurofibromatosis, Raised intracranial tension.

Giant pigmented nevi are infrequent. These cause social embarrassment and may undergo malignant transformation in upto 13.7% of cases.[1] These may also occur in 5% cases of neurofibromatosis. A giant pigmented nevus in the bathing trunk area is known as a bathing trunk nevus. Its association with neurofibromatosis is very rare.[2,3]

Case Report

A 17-year-old boy presented with bursting headache and vomiting for 3 months, difficulty in walking for 1 month and drowsiness for one week. He also had black hairy patches on his back, thighs, arms and face since birth. Hairs on these patches became coarse and dark black by age 11. Multiple, asymptomatic, small swellings appeared on the patch of the back at the age of 1 year and gradually increased in size. The boy was born of a full-term normal delivery. His milestones and sexual development were normal. His two younger sisters and parents did not have any apparent illness.

His pulse was 84 and BP 112/70. There was no fever, jaundice or lymphadenopathy. Chest, heart and abdomen were normal. He was drowsy, his speech was normal and gait was unsteady. He had bilateral papilloedema. Other cranial nerves were normal. There was no gross sensory-motor deficit. Plantar responses were extensor. Meningeal signs were negative. There was a large pigmented patch over the bathing trunk area extending from the scapulae to the buttocks and encroaching on the pubic area in front (Figs. 1 and 2). It was dark bluish-black and covered in the lower two-thirds with thick black 3-4 cm long hairs. There were 9 other similar though smaller (1 cm × 1 cm to 7 cm × 4 cm) patches with coarse hairs on the right arm, right thigh, right hypochondrium, forehead and face. Multiple swellings were present on the large patch on the back. Two swellings on either side were very large, hanging on pedicles and 12 cm × 10 cm to 25 cm × 17 cm in size. These were oval and soft in consistency. There were seven other hemispherical swellings—3 cm × 3 cm to 6 cm × 7 cm in size and soft to firm in consistency. One was cystic but with

Fig. 1. Bathing trunk nevus with hair.

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Fig. 2. Neurofibromatous tumours.

no sign of inflammation. The skin over the large swellings had normal colour, telangiectasia and sparse hair.

Haematological investigations and the chest, skull and spine X-rays were normal. The biopsy from the largest swelling revealed neurofibromatosis and that from the nevus on the back showed changes of pigmented hairy nevus with no melanomatous transformation. The patient did not allow more biopsies from other lesions. His headache, vomiting and drowsiness responded to intravenous mannitol and oral glycerol. Computed tomography of brain was planned but he was lost to follow up.

Comments

Large nevi may rarely be associated with melanocytosis of the central nervous system including meninges, brain and spinal cord, and can lead to raised intracranial tension and hydrocephalus. Malignant change has been reported in the meningeal infiltration.[4] Our patient had raised intracranial tension. There was no clinical or histopathological evidence of malignant change in the nevus. Hence, the possibility of intracranial metastasis is unlikely. The possibility of intracranial melanocytosis was seriously considered but we could not exclude an intracranial tumour in the absence of a computed tomography scan.

References

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