Cutis vertices gyrata (CVG) is characterized by thickening of the scalp, which becomes raised to form ridges and furrows resembling the cerebral gyri, which cannot be flattened by traction or pressure. The diagnosis is based on clinical features, and further investigations are needed to demonstrate the precise typical features. We describe how three-dimensional magnetic resonance imaging (3D MRI) can be used to visualize the extent of the lesions in primary non-essential CVG.

CASE REPORT

A 24-year-old man presented with deep furrows and convolutions on his forehead and scalp. Transverse furrows and convolutions on his forehead had developed 10 years earlier and vertical folds developed on the scalp 2 years later. The folds became deeper and increased in size to form ridges and furrows.

His medical history was not significant. He had no history of any disorders or inflammatory conditions of his skin or scalp besides the keloids on his anterior chest and both shoulders. He denied a family history of similar scalp condition.

Physical examination showed folds and furrows running transversely on the forehead (Fig. 1A) and anteroposteriorly on the scalp (Fig. 1B), which could not be corrected by pressure or traction. Laboratory examinations, such as complete blood-cell count, chemistry, venereal disease research laboratory (VDRL) test, urinalysis and chromosomal analysis, were all negative or normal. He had an amblyopia on his right eye which had gradually progressed and had conduction defect in the right visual pathway anterior to the chiasm by visual-evoked potential test. He had borderline intelligence on the Korean Wechsler Intelligence Scale. Pure-tone audiogram and skull X-ray were normal. Brain MRI revealed ischaemic changes: gliosis of both parietal lobes and periventricular leukomalacia of the left parietal lobe. A scalp skin biopsy showed an essentially normal histology. 3D MRI showed typical ridges and furrows of CVG on his forehead and scalp more clearly (Fig. 2).

DISCUSSION

CVG may be classified into primary and secondary forms (1). In primary CVG, ridges and furrows usually occur after puberty and show symmetrical distribution. A skin biopsy specimen reveals a normal appearance or thickened connective tissue, hyperplasia of appendages. The primary type can be further divided into essential and non-essential forms. The former may exist as a
solitary finding, with no association with neurological and ophthalmological disease and the latter may be associated with mental retardation, epilepsy and other brain or ophthalmologic abnormalities (1–4). Secondary CVG may appear at any age, skin folds are usually asymmetric and histology reveals various alterations depending on underlying causes such as tumours, neurofibromas, cerebriform intradermal nevi and inflammatory conditions. Systemic disorders associated with secondary CVG include acromegaly, myxedema, amyloidosis or pachydermoperiostosis (1–4).

The diagnosis of CVG can be made clinically, but the length and styling of hair makes it difficult to detect the characteristic features of CVG. A few reports of CVG used computed tomography or MRI as a diagnostic method, however, it is difficult to reveal the entire pattern of the scalp lesions by these methods (5–7). We found 3D MRI of the head could show the characteristic furrows and ridges of CVG more obviously.

REFERENCES