A 42 year lady presented with complaints of headache and vomiting for 10 days, followed by gradual deterioration in her consciousness for past 2 days. There was no history of fever or seizures. On examination her GCS was E4 M5 V2, vitals were stable and signs of meningeal irritation were absent, pupils were normal in size and reaction, plantar were bilaterally flexor and there was no focal neurological deficit. Rest of the systemic examination was within normal limit. Routine investigations revealed normal haemogram, blood sugar, liver and kidney function tests. Elisa for HIV 1 and 2 was non reactive. Contrast enhanced computed tomography (CECT) head was done which showed a heart shaped midline hypodense lesion involving the genu of corpus callosum and bilateral frontal lobes. There was gross perilesional oedema causing mass effect in the form of effacement of adjacent sulci and bilateral frontal horns (Figure 1). MRI brain showed a heart shaped hyperintense lesion of the genu of corpus callosum with oedema in bilateral frontal lobes on T2 weighted images (Figure 2). Contrast enhanced MRI brain revealed heterogeneous peripheral enhancement of the heart shaped lesion (Figure 3). CECT and MRI brain were suggestive of butterfly glioma. Patient was referred to neurosurgery department for biopsy and further management but...
patient attendant refused for any intervention and went home.

The differential diagnosis of mass lesion in corpus callosum includes butterfly glioma and rarely primary cerebral lymphoma, Toxoplasmosis, cerebral metastases and demyelinating butterfly pseudoglioma and neuronal ceroid-lipofuscinosis (Kufs’ disease). The ‘butterfly glioma’ refers to a high grade astrocytoma, usually a Glioblastoma multiforme (WHO grade IV), which crosses the midline via the corpus callosum. Other white matter commissures are also occasionally involved. Most frequently butterfly gliomas occur in the frontal lobes, crossing via the genu of the corpus callosum, however posterior butterflies are also encountered.

We have presented this Pictorial CME because of unusual radiological appearance to make physicians aware that whenever such type of bilateral involvement of white matter tract occurs especially in immunocompetent patient, possibility of high grade astrocytoma should be considered.

References


Hypomelanosis of Ito
Shrikant Sharma*, Ritesh Gupta**, Ganesh Narayan Saxena***, MS Raghu****, Anshul Patodia*****

Fig. 1 : Hypomelanotic patch (Ipsilateral to hypertrophy) (Streak type)

Fig. 2 : Hemihypertrophy (Right foot)

Fig. 3 : Hemihypertrophy Right lower limb

A 15 year old boy had recurrent seizure episodes at presentation with history of first seizure 5 years back. On examination he had hypopigmented lesions at forearm (Figure 1) and right thigh. Both sites showed streaks of hypopigmentation. He had coarse facies and divergent squint. The right arm and leg were bigger than that on the left side (Figures 2 and 3). His IQ score was 73. Routine blood investigations including blood sugar, serum electrolytes and calcium were normal. He had a history of surgery for right inguinal hernia. Brain MRI did not reveal any abnormality. Patient was on sodium valproate 600 mg/
day at presentation. Clobazem (10 mg/day) followed by phenytoin sodium (200 mg/day) were added subsequently to control seizures.

Hypomelanosis of Ito is thought to be third most common neurocutaneous disorder after neurofibromatosis type 1, and tuberous sclerosis. Syndrome is characterised with hypopigmented lesions that have a peculiar pattern of whorls, swirls, streaks, and patches. Previously known as incontinentia pigmenti acromians, was described by Ito in year 1952 with the clinical presentation of varying combination of abnormalities including central nervous system, eye, skeleton, and skin. The female predominance seen among HI patient points towards an X inactivation to become functional mosaics.

The cutaneous hypopigmentation is the only constant feature and best seen in dark skinned individuals as they have a unique distribution and appearance. They result from decrease in the number of melanocytes as well as of the number and size of melanosomes. Two clones of cells are randomly distributed in the primary streak in early embryogenesis. They migrate dorso-ventrally, proliferate and produce two populations of melanocytes with different pigment producing potential. The path of this migration are known as Blaschko lines.

Common manifestation of central nervous system involvement include mental retardation, seizures, language disabilities, and motor system dysfunction. Abnormalities seen on brain imaging include hemimegalencephaly, migration abnormalities, agenesis of corpus callosum, asymmetrical or symmetrical ventricular dilatation, focal cerebral atrophy with porencephalic ventricular dilation, hemi atrophy, diffuse cerebral atrophy, cerebellar hypoplasia or atrophy, vascular abnormalities or rarely tumour.

Ocular abnormalities include heterochromic iris, strabismus, myopia, opaque cornea, optic atrophy and microophthalmia.

Other associated anomalies include macrocephaly microcephaly, hemihypertrophy, kyphoscoliosis, coarse facies, genital anomalies, inguinal hernia, congenital heart diseases, hypertelorism, cleft palate and abnormalities of teeth and feet and cardiac and genital anomalies.

No universally accepted diagnostic criteria for HI exist. There is no treatment for disorder, except for symptomatic management. The lack of published series on the natural history of disease makes accurate prognosis difficult.

References
2. Sybert VP. Hypomelanosis of Ito: a description, not a diagnosis. J invest Dermatol 1994;103(5-suppl);141S-143S.
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