Complex limbal choristoma in linear nevus sebaceous syndrome managed with scleral grafting

Nitin Trivedi, Girish Nehete

Linear nevus sebaceous syndrome (LNSS) is characterized by nevus sebaceous, mental retardation, seizures, and ocular abnormalities such as complex limbal choistoma. A young male with history of mass in right eye and blackish discoloration of skin over right and left side of forehead since birth presented with foreign body sensation and diminished vision in right eye. Ocular examination showed mass over epibulbar region with chorioretinal coloboma and posterior staphyloma in right eye and megalocornea in left eye. Histopathology report revealed complex limbal choristoma with compound melanocytic nevus. The case was managed by surgical excision of the limbal mass and filling the gap with scleral graft.

Key words: Chorioretinal coloboma, complex limbal choristoma, nevus sebaceous

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Linear nevus sebaceous syndrome (LNSS) is characterized by nevus sebaceous, mental retardation, seizures, and ocular abnormalities. Epibulbar choristomas are common, while posterior scleral choristomas are very rare.[1] We report the clinical and histopathological findings and surgical management of a patient with LNSS.

Case Report

A 16-year-old male came to our outpatient clinic with complaints of foreign body sensation and diminished vision in right eye. He gave history of blackish discoloration of the skin over right and left side of forehead since birth. There was no history of mental retardation and convulsion [Fig. 1].

Birth history was not significant and there was no family history of consanguinity.

General examination showed melanotic patches over right and left side of face, forehead, and temple. The scalp region also showed involvement in the form of nevus sebaceous in its characteristic clinical appearance and patchy alopecia.

Magnetic resonance imaging (MRI) of brain was normal.

Examination of right eye showed visual acuity up to 2 m. Finger counting and the slit lamp examination showed a large mass hanging over temporal bulbar conjunctiva involving superotemporal quadrant of the cornea [Fig. 2]. Fundus examination showed posterior staphyloma, chorioretinal atrophic patches, and coloboma over superotemporal arcade.

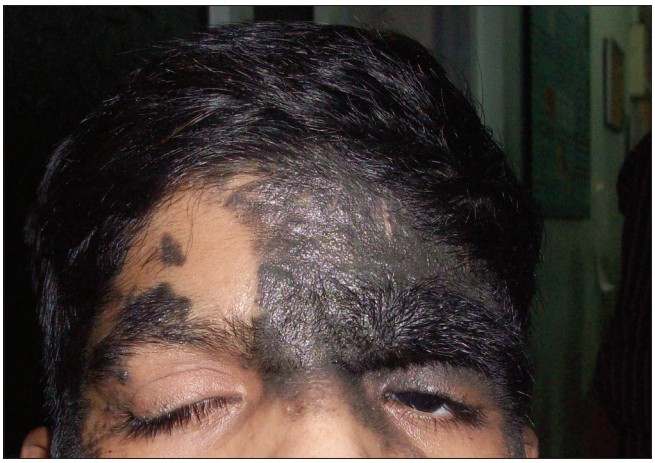

Figure 1: Photograph of a patient

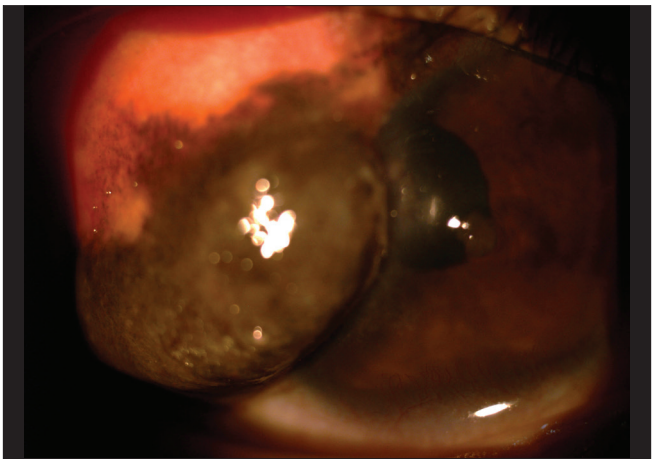

Figure 2: Slit lamp photograph of right eye showing limbal mass

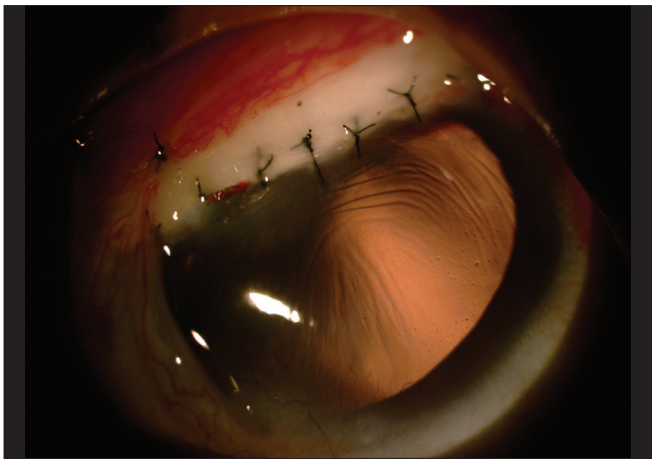

Figure 3: Postoperative Slit lamp photograph of right eye with sclera graft showing red fundal glow

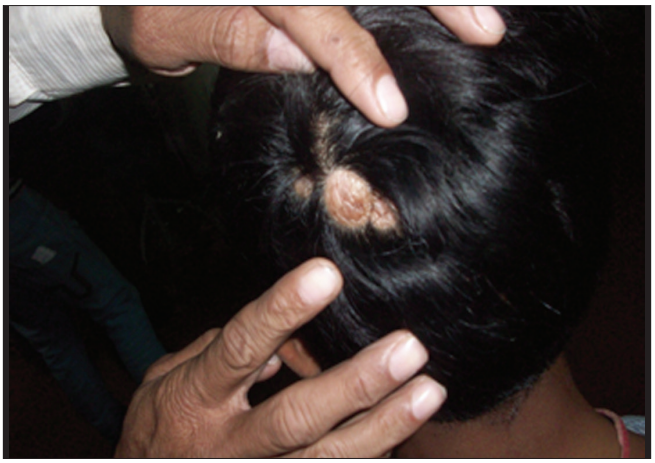

Figure 4: Scalp involvement in the form of nevus sebaceous and patchy alopecia

Examination of left eye showed visual acuity 20/40, megalocornea, and nevus over temporal limbus with Intraocular pressure of 18.9 mmHg and fundus was normal.

Ultrasonography preoperatively showed both eye axial length 26 mm, only right eye showing posterior staphyloma, rest no abnormality detected. Surgical excision of the limbal mass was planned [Fig. 3].

An attempt was made to remove the limbal mass surgically under peribulbar anesthesia. A fine dissection was carried out to separate the mass from sclera but separation was not possible and as the dissection went deeper, uveal tissue was visible. The mass was found to be involving superotemporal quadrant of the cornea. At the end of dissection, mass was separated leaving behind a gap of size 1 × 1.5 cm in the scleral coat showing black uveal tissue. The excised mass consisted of pigmented and nonpigmented areas and was soft to hard in consistency which was sent for histopathological examination. Preserved sclera of size 1 × 1.5 cm was fashioned and sutured to cover up the defect with 6-0 black nylon in intermittent fashion.

Postoperatively, fundal glow improved but improvement in visual acuity was negligible due to chorioretinal coloboma.

Gross/macroscopic examination showed grayish black mass measuring 1 × 0.5 cm and another mass of brownish color measuring 1.5 × 1 cm and mature cartilage measuring 1.5 × 2 cm.

Microscopic examination of grayish black mass showed stratified squamous epithelium with adnexal structure, lacrimal glands, adipose tissue, neural tissue, and mature cartilage. Another brownish colored mass revealed area of stratified squamous epithelium with compound melanocytic proliferation, pigmentation without atypia and mitosis, suggestive of compound melanocytic nevus.

Histopathological examination was consistent with complex ocular choristoma.

Discussion

Epibulbar choristomas are common in LNSS and may occur bilaterally in some cases. Complex choristomas are rare epibulbar tumors and are characterized by the presence of two or more ectopic tissues. Surgical treatment may be required because of involvement of the cornea. In our case, the choristoma was unilateral and histopathologically complex, showing stratified squamous epithelium with adnexal

structure, lacrimal glands, adipose tissue, neural tissue, and mature cartilage.[2,3] The choristoma in the right eye was excised and scleral grafting was performed as choristoma was involving more of scleral side. Other ocular findings which may be present with LNSS are eyelid colobomas and masses, ptosis, nystagmus, strabismus, corneal vascularization, microphthalmia, macrophthalmia, iris coloboma, cataract, chorioretinal coloboma, staphyloma, optic disc coloboma, optic nerve glioma, pseudopapilloedema, choroidal osteoma, and cortical visual impairment. Our case had unilateral limbal choristoma, posterior staphyloma, and chorioretinal coloboma over superotemporal arcade and left eye showed megalocornea and nevus over temporal limbus. Also compound melanocytic nevus involving periocular region of face was noted which was purely coincidental finding.

Nevus sebaceous may be associated with malignancy. Therefore, these patients must be monitored closely. In our patient, the nevus sebaceous has shown no evidence of malignant transformation till 1 year of follow-up examinations. Patchy alopecia was also observed in our case [Fig. 4]. Neurological abnormalities, seizures and mental retardation, quadriparesis, microcephaly, and diencephalic syndrome may be observed in LNSS patients. But, our patient had no history of developmental delay and convulsion and no evidence of mental retardation.

Corneal grafting was tried in these patients but no encouraging results found due to graft vascularization.[4] Amniotic membrane transplant was also tried after excision of complex limbal choristoma for surface reconstruction.[5] Complex choristomas are usually deep and scleral grafting may be required to achieve the anatomical integrity. We have done scleral graft in our patient, which has proved to be promising.

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