anomalies, it is characteristically accompanied by ipsilateral heminasal hypoplasia or aplasia and rarely by choanal atresia [7]. Ocular and/or ocular adnexal (anophthalmia, microphthalmia, microcornea, lenticular opacities, cyclopean eye, and colobomas of the choroid, retina, iris, and eyelids) ndings as well as cle lip and/or palate are the most common anomalies seen in conjunction with PL and have been used as the basis for a classi cation system [8,9]. Group 1 consists of isolated PL without other associated anomalies (9%). Group 2 consists of PL with associated ipsilateral nasal defect (23%). Group 3 consists of PL with associated ipsilateral nasal and ocular and/or ocular adnexal defect (47%). Group 4 includes the features of group 3 with the addition of cle lip and/or palate (21%). However Sakamoto, et al. rede ned classi cation and added two new groups including group 5 as hypertelorism with encephalocele and group 6 as hypotelorism [10].

Although most patients with proboscis lateralis do not have serious central nervous system abnormalities, proboscis lateralis may coexist with central nervous system anomalies and early neuro-imaging is indicated to rule out intracranial abnormalities [11].

Initial reports regarding the treatment of PL recommended simple surgical excision of the proboscis. More recently, surgical management of PL has been approached with reconstruction in mind. Because there is some variability in facial anomalies and the degree of nasal hypoplasia seen with proboscis lateralis, management must be individualized [4,5]. Due to the variety of maxillofacial and ocular disease seen with proboscis lateralis, optimal care of the patient warrants a multidisciplinary approach that may involve an otolaryngologist or oromaxillofacial surgeon, plastic surgeon, and ophthalmologist.

Conclusion

PL is a rare congenital anomaly with a characteristic appearance. Computed tomography and MRI are complementary in determining the extent of the bony and so tissue components of the anomaly. Proboscis lateralis might be associated with developmental anomalies of the ipsilateral nasal and ocular structures. Complete surgical excision at the base of the proboscis is desirable as a primary procedure if there ostT2j /T1_ia185j 0.-5