Chapter 13 Chiari Malformation



These may or may not be accompanied by syringomyelia. These can also rarely be associated with Ehlers Danlos syndrome (EDS). Often Chiari 1 malformations are asymptomatic and can then be managed with observation.

For Chiari 1 malformations that are causing significant symptoms, posterior decompressive surgery is appropriate. The MIST (minimally invasive subpial tonsillectomy) procedure [86] is an excellent option when surgery is needed. It involves a smaller incision, a smaller opening of bone (occiput just 2 cm from the foramen, with just part of the C1 lamina), a linear dural incision, and resection of the cerebellar tonsils. Alternatively, "shrinkage" of the tonsils with a low setting on the cautery system may also be adequate (see Fig. 13.1). Care must be taken not to injure the PICA artery loops or the spinal accessory nerves. Excessively large bone work far from the foramen magnum is not clearly helpful. And while suturing in a large dural patch graft does provide extra intradural room, the patch graft adds time and complexity to the procedure that is not clearly necessary to decompress the foramen magnum and makes pseudomeningoceles and CSF leaks much more likely. Of note, if a Chiari malformation is felt to be secondary to some other condition, such as idiopathic intracranial hypertension, it is usually best to address that primary condition first.



Fig. 13.1 This is a 23-year-old woman with persistent bothersome occipital headaches worse with coughing and straining. MRI showed a Chiari 1 malformation (**a**: sagittal T1 MRI image). The patient underwent suboccipital craniectomy and cerebellar tonsillectomy (MIST procedure). Her pre-operative symptoms resolved. Postoperative MRI showed resection/decompression of cerebellar tonsils (**b**: sagittal T1 MRI image)