Late-onset progressive facial hemiatrophy (Parry-Romberg syndrome)

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A 43-year-old woman was referred for a computed tomography (CT) scan of her midface due to a progressively evolving right facial hemiatrophy. Though it began ten years earlier it became stable for the past one year. The patient’s medical history and family records were non-contributory, with no reports of trauma, use of medication or major illness. Her neurological examination was normal. On physical examination, there was remarkable wasting of soft tissues on the right side of her face, with thinning of skin and subcutaneous fat, scant eyebrows and lashes and prominence of the zygomatic arch. No discoloration or textural changes of skin were noticed. Right enophthalmus was also seen, with mild homolateral ptosis of the upper eyelid. A previous non-enhanced CT scan of the brain was normal.

Discussion

Initially described by Parry in 1825 and systematized by Romberg in 1846, progressive facial hemiatrophy syndrome is a controversial and poorly understood entity, with sporadic occurrence and no known mendelian inheritance. It is a slowly progressive disorder, affecting chiefly females in their first and second decades of life, characterized by unilateral atrophy of facial tissues, mostly skin and subcutaneous fat and, in a lesser degree, muscles and bones. It is closely associated with linear scleroderma en coup de sabre. However, both conditions may be overlapping and there may be evolution from one process into another. Its origin is unknown, and several hypotheses have been proposed (encephalitis, trauma, scleroderma, migraine, infection, vasculitis, genetic factors, malformative hypothesis), but a multifactorial pathogenesis may be most likely. Protein multi-systemic manifestations have been described, including ocular involvement, ear, nose and throat disorders and neurological findings.

Previous reports of Parry-Romberg syndrome in imaging literature have emphasized neuroimaging findings which are variable and non-specific. Increased signal in the white matter on long-TR sequences (which tends to progress to encephalomalacia), meningeal enhancement, intracranial calcifications, central cerebral atrophy, infarctions in the corpus callosum and cortical thickening have all been described. Although CT scan was regarded as normal in this patient, magnetic resonance imaging (MRI) was not performed and perhaps some additional findings could have come to light after the MRI.

Imaging findings of facial changes in this entity are not largely described, probably because they are clinically obvious, unlike...
Figure 2: Axial computed tomography images from below to above, with bone settings, revealing asymmetry of maxillary sinuses, being the right one slightly smaller. Zygomatic arches are roughly symmetric, as well as orbital walls.

Figure 3: Three-dimensional reconstruction with bone algorithm showing symmetry of orbits and zygomatic arches. Right side of the maxilla is somewhat depressed.

Figure 4: Three-dimensional reconstruction with soft-tissue algorithm demonstrating important wasting of soft tissues of the right side of the face. The zygomatic arch protrudes under the skin at right.

CNS alterations. According to previous reports, our patient had facial atrophy predominantly at the expense of skin and subcutaneous fat, with lesser involvement of bone and muscles. Our patient started her disease when she was in her forties so her craniofacial development was already complete. That is why her facial bones showed no significant asymmetry even after ten years of evolution, in spite of severe atrophy of overlying soft tissues.

In conclusion, although a disease of children and teenagers, the Parry-Romberg syndrome may occasionally occur in adults. The severity of the neurological findings and of the facial asymmetry in these cases may be lesser than when this entity has an early onset, probably because the central nervous system and the craniofacial structures are already fully developed.

References