Masayuki Shintaku · Kenichiro Maeno · Hidetoshi Okabe

Chondroid chordoma of the skull base: immunohistochemical and ultrastructural study of two cases with special reference to microtubules within rough-surfaced endoplasmic reticulum

Abstract  Two cases of skull base chordoma (case 1, a 57-year-old woman; case 2, a 69-year-old woman) were investigated immunohistochemically and ultrastructurally. The tumors showed histopathological features typical of chondroid chordoma and contained both classical chordomatous and hyaline cartilaginous components. Tumor cells were immunoreactive for cytokeratin, vimentin, and S-100 protein, but negative for microtubule-associated protein 2 and class III beta-tubulin (tub-B3). Tumor cells of case 2 were immunoreactive for tau-protein and class II beta-tubulin (tub-B2), whereas those of case 1 were negative. Ultrastructurally, tumor cells in both cases showed the presence of abundant glycogen granules, well-developed intracellular organelles, and desmosome-like junctions. In case 2, several microtubules were closely packed and ran parallel or in random directions within the dilated cisterns of rough-surfaced endoplasmic reticulum (rough ER). “Microtubules within rough ER” has been described in several neoplasms, including classical and chondroid chordomas. Although previous reports documented the tub-B3 immunoreactivity in chordomas, our results suggested that, in our case 2, the predominant isoform of beta-tubulin in microtubules within rough ER was not tub-B3 but tub-B2.

Key words  Chondroid chordoma · Skull base · Immunohistochemistry · Ultrastructure · Microtubules · Rough-surfaced endoplasmic reticulum · Class II beta-tubulin

Introduction

Chondroid chordoma is a variant of chordoma that arises almost exclusively in the clival region of the skull base and shows relatively favorable prognosis in comparison with classical chordoma. Histopathologically, chondroid chordoma contains a significant amount of hyaline cartilaginous matrix in addition to the chordomatous component. Since the first description of this variant by Heffelfinger et al., many cases have been reported, and the ultrastructural characteristics of this neoplasm have also been described by a few investigators.

We examined two cases of chondroid chordoma of the skull base. In this article, we report the histopathological, immunohistochemical, and ultrastructural findings of these cases, with special reference to “microtubules within rough-surfaced endoplasmic reticulum (rough ER),” which was found in case 2. Tumor cells in case 2 were immunoreactive for tau-protein and class II beta-tubulin, whereas those in case 1 were not immunoreactive for these two proteins. This immunohistochemical finding appeared to correspond with the presence of microtubules within rough ER in case 2.

Clinical histories

Case 1

The patient was a 57-year-old woman who presented with visual disturbance. Ophthalmologic examination demonstrated bitemporal hemianopsia, and cranial magnetic resonance imaging (MRI) disclosed a partly calcified tumor measuring 4 cm in diameter in the clival region. It compressed the cranial nerves and invaded the sphenoid and ethmoid sinuses. The tumor was extirpated subtotally by a transsphenoidal approach, and the subjective symptoms were improved remarkably. Postoperative MRI suggested the persistence of a small amount of residual tumor, and gamma-knife therapy is now scheduled as a treatment modality.

Case 2

The patient was a 69-year-old woman who presented with vertigo of acute onset after a floating sensation. Cranial
Fig. 1. The tumors of both cases showed a trabecular or sheet-like, solid growth of large polygonal cells. Some individual tumor cells surrounded by lacunae were embedded within the hyaline cartilaginous matrix. a Case 1; b case 2. Hematoxylin and eosin (H&E) stain

Fig. 2. Tumor cells had round nuclei and abundant pale cytoplasm that contained occasionally single or multiple vacuoles (“physaliphorous cells”). Some tumor cells showed a “ground-glass” appearance of the cytoplasm. a Case 1; b case 2. H&E stain

Fig. 3. The cytoplasm of tumor cells was strongly and diffusely immunoreactive for cytokeratin (a) and S-100 protein (b). Tumor cells in case 1 were negative for class II beta-tubulin (tub-B2) (c), but those in case 2 were diffusely immunoreactive for tub-B2 (d). a, c Case 1; b, d case 2. Labeled streptavidin–biotin method