# Case Report Pathological features of myxopapillary ependymomas in lumbar spinal canal: report of two cases

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**Abstract:** Myxopapillary ependymoma almost only occurs in conus medullaris, cauda equine and terminal filament, and myxopapillary ependymoma has distinctive morphology and immunohistochemical phenotype. The article will discuss two rare cases report of myxopapillary ependymoma. The two cases occur in lumbar spinal canal, tumor cells arrange in well-differentiated blood vessels or around the mesenchymal axis of connective tissue without cells with the nipple radial structure, and have obvious micro-capsule structure. There are accumulated with lots of grume between blood vessels and tumor cells. And are abundant with interstitial blood vessels, and bleeding, collagen in focal mesenchyme, and hemosiderin deposition in small focus. Immunohistochemical markers, including GFAP, Vimentin, S-100, NSE, showed positive expression in tumor cells, AE1/AE3, EMA, Syn, CD34, D2-40, showed negative expression in tumor cells, the proliferation index of Ki-67 was less than 1%. And we will analysis the clinicopathological features, and diagnosis differential diagnostic of the lumbar spinal canal of Myxopapillary ependymomas.

Keywords: Myxopapillary ependymoma, lumbar spinal canal, differential diagnosis, immunohistochemistry

#### Introduction

Myxopapillary ependymoma (MPE) is a rare subtype in ependymoma. In 1932, Kernohan et al described for the first time the morphological characteristics and biological behaviors of the tumor. This tumor almost only occurs in conus medullaris, cauda equine, and terminal filament and especially occurs in young people. Morbidity is higher in male than in female. The tumor is slow growth, but maybe has local recurrence or metastases. WHO grading is Grade I. We reported 2 cases with MPE and discused its clinic-pathologic features, immunophenotype and differential diagnosis.

#### Case report

Case 1: a 24-years old male presented to the third affiliated hospital of Soochow University suffering from lumbo-sacral pains for approximately four years. The patient appeared lumbosacral pains four years ago, and the pains was ache, intermittent, and improved after taking a break. Symptoms after attacked repeatedly. In recent one year, lumbo-sacral portion pains have been exacerbated, accompanying with pains in bilateral hips and back thigh. Walking and standing for a long time, pains were exacerbated. The computed tomography (CT) scan of the chest showed lumbar spine was oval softtissue density structure in L2/3 spinal canal. Dural sac was pressed slightly. Bilateral recess was stenosis.

Case 2: female, 34 years, suffering from lumbosacral pains with numbness and ache for more than 3 months. The patient had lumbago with radiating pains of right lower limb and skin numbness of right acrotarsium under no obvious predisposing causes 3 months ago. Walking and standing for a long time, pains were exacerbated; the symptom might be alleviated after lying. The computed tomography scan of the chest showed lumbar spine was flake and high density shadows in L2 spinal canal. Physiological curvature of lumbar spine was complete and the centrums bone had no obvious hyperostosis. Soft-tissue density shadows around the L3/4, L4/5 and L5/S1 centrums were bulged slightly. Dural sac was pressed, and bilateral recess was stenosis (Figure 1).



Figure 1. A: CT scan of the chest showed flake and slightly hypo-dense shadows in L2 spinal canal; B: Soft-tissue density shadows around the L3/4, L4/5 and L5/S1 centrums and slight bulged.

## Macro examination

The two cases stripping tumor tissues were broken, parts of tissues were lobulated or botuliform with envelope. The section was off-white or off-red and tender, accompanying with bleeding.

## Microscopic examination

Optical microscopy revealed cells arranged well-differentiated blood vessels or around the mesenchymal axis of connective tissue without cells with radial structure of papilla, and had obvious micro-capsule structure. Cells covering on the surface of papillary structure arranged regularly, had clear boundary, which often was monolayer, and also can be multiple layers. Its protuberance pointed to the center, cell nucleus located in the periphery. Micro-capsule structure had abundant grume. Tumor cells revealed flat, cubic or short fusiform. The cytoplasm was middle, eosinophilic red or transparent with vacuolization. Nucleus was round or short fusiform, and nuclear membranes were obvious, chromatin was fine and visible small nucleolus. A large amount of mucus accumulated between blood vessels and tumor cells. Mesenchymal blood vessels were abundant, and visible cavernous hemangioma area, parts of blood vessels were hyaline degeneration, and bleeding and collagen in mesenchyme, and

hemosiderin deposition in small focus. Tumor cells morphology and size were consistent, no obvious atypia and polymorphism. Nuclear mitotic occasionally appeared or no, and necrosis (**Figure 2**).

# Immunohistochemistry

The tumor cells were stained positive for GFAP, S-100, NSE, but were negative for AE1/AE3, EMA, Syn, CD34, D2-40. The proliferation index of Ki-67 was less than 1%. AB-PAS specific stain displayed blue grume (**Figure 3**).

# Treatment and prognosis

After complete excision of tumors, and no radiotherapy, the first patient had no recurrence at one-year follow-up. The second patient just conducted complete excision of tumors, follow-up were still needed to further evaluate the effects.

# Discussion

Myxopapillary ependymoma (MPE) is a rare subtype in ependymoma [1]. In 1932, Kernohan et al described for the first time the morphological characteristics and biological behaviors of the tumor [2]. And it almost only occurs in conus medullaris, cauda equine and terminal filament. Extradural MPE is rare. According to related literatures, the primarily MPE occurs



**Figure 2.** A: Large amount of mucus accumulated between blood vessels and tumor cells, obvious micro-capsule structure; B: Tumor cells reveal papillary structure; C: Cavernous hemangioma; D: Abundant mesenchymal blood vessels, hyaline degeneration of blood vessels (magnification, ×100).

in tail sacrum, front sacrum, soft-tissue close sacrum and broad ligament; maybe originate from extradural residual terminal filament or nervi duct tail-end remnants [3]. The cases mainly come from the ependymocyte in the central canal of terminal filament. MPE is a sluggish tumor. Surgery was the first choice for treatment, and surgical methods included gross total removal, piecemeal total removal and subtotal removal. But, the surgical may result in planting metastasis tumor cells [4]. There was no systematic molecular study for MPE, currently. Some research found that MPE have the multiploidy for chromosome 7 and characteristic increase in copy number for chromosome 7, may be connected with pathogenesis of MPE.

## Differential diagnosis

Chordoma: It is a low or moderate malignant tumor occurred in sacrococcygeal region. Tumor cells often destroy the sacrum and forms characteristic bileaflet structure, and lamina septate fiber. Tumor cells reveal funicular, nested or unicellular floating in the light-blue grume matrix, and vacuolated cells of different sizes. Small cells are like vacuole-type signet-ringcell, the big are like balloon-type cavity [5]. Generally, it doesn't form the papillary structure, AE1/AE3, EMA and S-100 proteins immunostaining were positive, whereas GFAP was negative [6].

Schwannoma: When the mucus papillary structure of MPE was not obvious and arranged with sarciniform of fusiform cells, it needed to identify Schwannoma. The fine structure of Schwannoma has complete capsulare and occurs in two forms: Type Antoni A is composed of Schwann cells whose nuclei are arranged in palisading rows with greatly attenuated cytoplasmic processes extending from the Schwann cells in parallel alignment; Antoni B is characterized by loosely arranged Schwann cells set in



**Figure 3.** A: Immuno-staining showing tumor cells strongly positive S-100; B: Positive for Vimentin; C: Positive for GFAP; D: Positive for NSE; E; Negative for CD34; F: AB-PAS reveal blue grume (magnification, ×100).

meshwork of macrocysts and reticular fibers. Sometimes, Schwann cells can form Verocay body, but it has no papillary structure. Moreover, Mucus accumulation in cystic cavity of Schwann cells. GFAP protein immunostaining was little positive or negative [7].

Spinal meningioma: Spinal meningioma cells arranged with beam cross or lobulated, and

revealed vortex structures and psammoma body. Spinal meningioma cells boundaries were unclear, nucleus was ovoid, chromatin was fine, and parts of spinal meningioma cells were fusiformis, like fibroblast. EMA protein immunostaining was positive [6, 8].

Digital papillary adenocarcinoma: Papillary adenocarcinoma cells atypias was obvious, vis-

ible nuclear division, complicated papillary and clear cells lumens. AE1/AE3 and EMA proteins immunostaining was positive [9].

Extra-adrenal paraganglioma: Extra-adrenal paraganglioma usually occurred in spinal dura mater of cauda equine and terminal filament, adhesive to cauda equine. Tumor epithelial cells were composed with chief cells and sustentacular cells around chief cells. Chief cells arranged with organic samples or cell ball-like structure, and revealed oval or polygon. Chief cells cytoplasms were eosinophilic, slight-granular and usually larger vesicular nucleus. Parts of extra-adrenal paraganglioma cells were acinar or adenoid. The NSE, Syn, CgA proteins of chief cell immunostaining was positive, whereas GFAP was negative. And the S-100 protein of fusiformis sustentacular cell was positive [10].

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#### Disclosure of conflict of interest

None.

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