

# Cerebellar Squamous Cell Carcinoma Due to Malignant Transformation of Cerebellopontine Angle Epidermoid Cyst, Report an Interesting Case and Review the Literature

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**Abstract:** Malignant transformation of an epidermoid tumour is a rare entity that in almost all patients occurs at the same site of the primary lesion. We report a case of an epidermoid tumour with malignant transformation to squamous cell carcinoma (SCC) at the adjacent site but without any relation to the primary site of the tumour. A 30-year-old patient with a history of cranial surgery and resection of cerebellopontine (CP) angle epidermoid cyst five years ago, presented with a headache, nausea, and vomiting. Physical examination showed no neurological deficit. The brain magnetic resonance imaging (MRI) demonstrated a well-defined lesion within left middle cerebellar peduncle with no relation to CP angle cistern (the previous tumour site). It was isointense on T1, isointense on T2 and had a rim enhancement on gadolinium (GD) injection. Via retrosigmoid and transcortical approach, total resection of the tumour was performed. During the surgery, there was no visible relationship between the current lesion and the previously resected lesion site. Histopathology revealed squamous cell carcinoma. The systemic survey to finding a probable origin of the tumour was negative and the patient referred for performing brain radiotherapy. We are reporting a case of malignant transformation of epidermoid cyst separate from primary location. Moreover, malignant transformation can occur years after index surgery even after gross total resection.

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## Introduction

The intracranial epidermoid tumour is a rare and benign tumour. Malignant transformation of an epidermoid tumour is a rare entity that in almost all patients occurs at the same site of the primary lesion (Lakhdar et al., 2011). Now we report a case of an epidermoid tumour with malignant transformation to squamous cell carcinoma (SCC) at the adjacent to the surgical site but without direct relation to the primary site of the tumour.

## Case report

A 30-year-old patient presented with left hemifacial spasm. The physical examination demonstrated decreased left side hearing loss, decreased sensation in V1 and V2 territory and deviation of the tongue to right and uvula to the left. The brain magnetic resonance imaging (MRI) revealed an extraaxial cystic mass in the left cerebellopontine (CP) angle compressing cerebellum, cerebellar peduncle and brain stem (Figure 1). With a diagnosis of an epidermoid tumour, retrosigmoid craniectomy and near-total resection of tumour was performed. Only a very small part of tumour adhesive to the facial nerve remained in place to prevent nerve damage. Histopathology confirmed the diagnosis of the epidermoid tumour. The patient had only short time follow-up postoperatively. About five years later, he returned because of a headache, nausea, and vomiting. Physical examination showed no neurological deficit. The brain MRI demonstrated a well-defined lesion within left middle cerebellar peduncle with no relation to CP angle cistern that was isointense

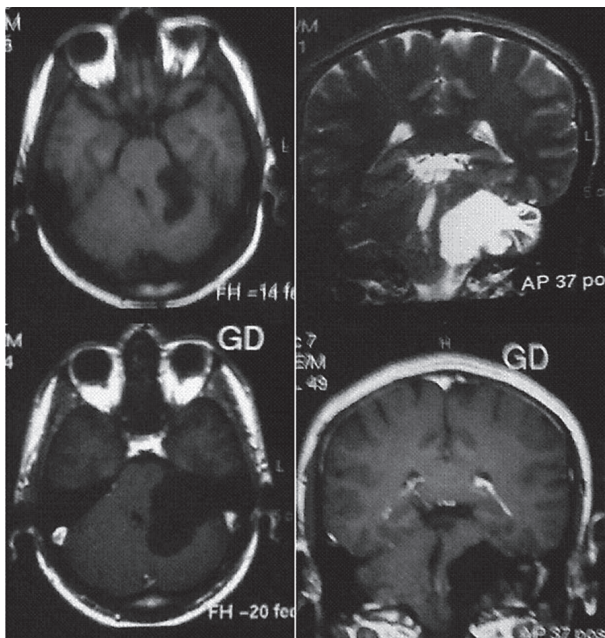


Figure 1 – The brain magnetic resonance imaging (T1, T2 and T1 with gadolinium injection) demonstrated an extraaxial cystic mass in cerebellopontine angle.

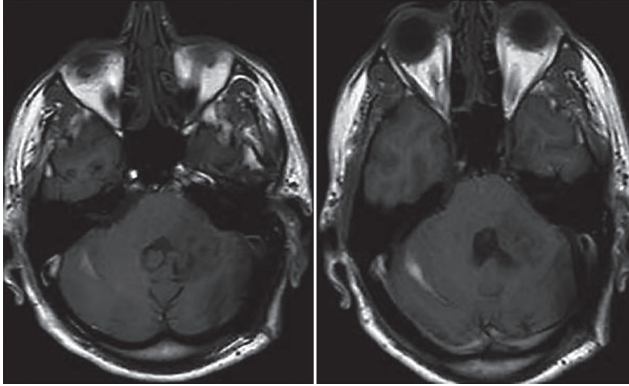


Figure 2 – The T1 images sequences of brain magnetic resonance imaging revealed the lesion.

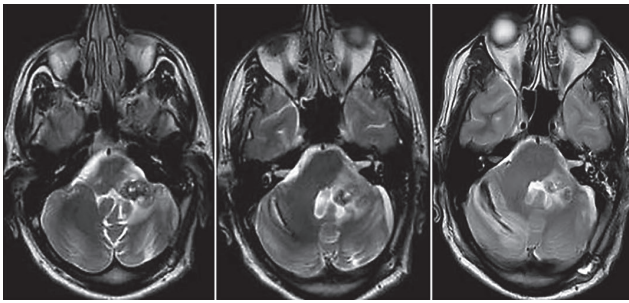


Figure 3 – The T2 images sequences of brain magnetic resonance imaging revealed the lesion within middle cerebellar peduncle.

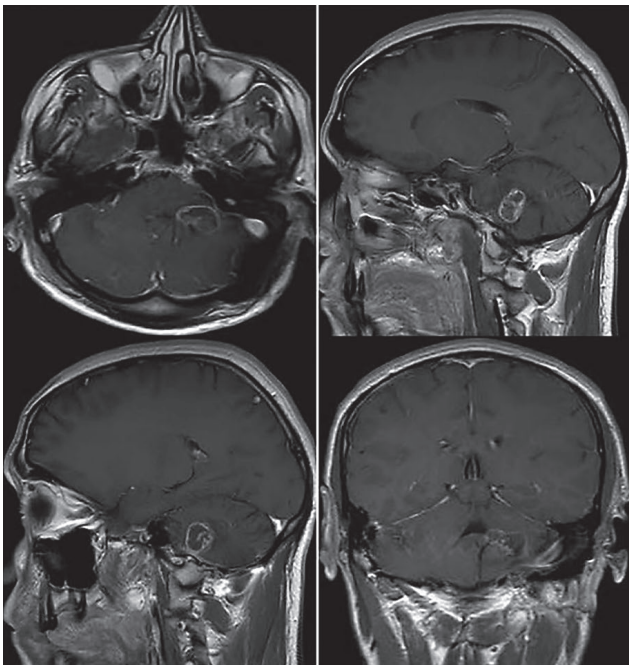


Figure 4 – The brain magnetic resonance imaging with gadolinium injection demonstrated the ring enhancement of the middle cerebellar peduncle lesion.

**Table 1 – Summarizes the most information of some studies have reported malignant transformation epidermoid cyst to squamous cell carcinoma**

Study	Sex	Age	Location	Interval to MT	Location of SCC	Final diagnosis	Treatment	Outcome	Follow-up
Hamlat et al. (2003)	F	54	parietotemporal	10 m	operation field	SCC transformed from the EC/ leptomeningeal carcinomatosis	-/IT chemotherapy	expire	16 m
Lakhdar et al. (2011)	M	50	CP angle	6 m	operation field	SCC transformed from the EC	surgery/ radiotherapy	good	3 m
Tamura et al. (2006)	F	56	CP angle	8 y	operation field	SCC transformed from the EC	surgery/ radiosurgery	good	4 m
Kodama et al. (2007)	M	67	CP angle	2 m	operation field	SCC transformed from the EC/ leptomeningeal carcinomatosis	surgery/ radiosurgery	expire	13 m
Kim and Kim (2008)	F	72	CP angle	2 m	operation field	SCC transformed from the EC	surgery/ radiotherapy	good	12 m
Chon et al. (2012)	M	43	CP angle	5 m	outside of operation field	SCC transformed from the EC	surgery/ radiosurgery	recurrence	2 y
Hao et al. (2010)	F	61	CP angle	6 y	MT over the primary lesion	SCC transformed from the EC	surgery/-	expire	36 d
Ge et al. (2009)	M	44	temporal lobe	6 y	operation field	SCC transformed from the EC	surgery/?	?	?
Nakao et al. (2010)	F	74	CP angle	20 y	operation field	SCC transformed from the EC	surgery/ radiotherapy	good	17 m
Kano et al. (2010)	F	64	left parapatent extension to medial temporal	14 y	operation field	SCC transformed from the EC	surgery/ radiotherapy	death	2 y
Nishiura et al. (1989)	M	38	CP angle	7 m	operation field	SCC transformed from the EC	surgery/ chemotherapy	alive	2 y
Murase et al. (1999)	F	50	CP angle	11 y	operation field	SCC transformed from the EC	surgery/ chemotherapy/ radiosurgery	good	5 y

Study	Sex	Age	Location	Interval to MT	Location of SCC	Final diagnosis	Treatment	Outcome	Follow-up
Knorr et al. (1991)	M	74	CP angle	13 m	operation field	SCC transformed from the EC	surgery/ radiotherapy	death	7 w
Feng et al. (2014)	M	42	CP angle	–	first presentation	malignant epidermoid cyst	surgery/ radiotherapy	good	6 m
Fox and South (1965)	M	43	anterior temporal	7 y	operation field	SCC transformed from the EC	surgery/–	death	1.5 m
Ozutemiz et al. (2017)	M	64	posterior horn of the left lateral ventricle	23 y	adjacent to tumour	SCC transformed from the EC	surgery/–	recurrence	3 m
Ding et al. (2016)	F	55	temporal region and prepontine area	7 m	operation field	SCC transformed from the EC	surgery/–	death	6 m
Nosaka et al. (1979)	M	46	CP angle	5 m	adjacent to tumour within brain stem	SCC transformed from the EC	–/–	death	2 m
Asahi et al. (2001)	F	55	CP angle	13 y	operation field	SCC transformed from the EC	surgery/–	death	3 m
Our study	M	30	CP angle	5 y	middle cerebellar peduncle	SCC transformed from the EC	surgery/ radiotherapy	good	1 y

SCC – squamous cell carcinoma; EC – epidermoid cyst; MT – malignant transformation; CP angle – cerebellopontine angle; IT – intrathecal; M – male; F – female; d – day; w – week; m – month; y – year

on T1, isointense on T2 and had a rim enhancement on gadolinium (GD) injection (Figures 2–4). The patient underwent total resection of the tumour via retrosigmoid and transcortical cerebellar hemispheric approach. During the surgery, there was no visible relationship between the current lesion and the previously resected lesion site. Histopathology revealed squamous cell carcinoma with the origin of the epidermoid tumour. The systemic survey to find a probable origin of the tumour was negative and the patient referred for performing brain radiotherapy. On last follow-up about 2 years later, the patient demonstrated no significant neurological sign and symptom, and no tumour recurrence.

## Discussion

The intracranial epidermoid tumour is a rare and benign tumour that has a slow growth and includes 0.2–1.8% all intracranial tumours (Tamura et al., 2006; Lakhdar et al., 2011). The remnant of the squamous epithelial include in the neural tube at the time of last separation phase between three to five weeks of fetus life (Hao et al., 2010). A malignant transformation from an epidermoid tumour is a rare entity and was reported for the first time by Ernst in 1912 (Lakhdar et al., 2011). There is controversy about the exact mechanism of malignant transformation of an epidermoid tumour, but inflammation due to reaction to a foreign body, *in situ* carcinoma, chronic inflammation due to frequent cyst rupture and subtotal resection cyst wall may be the reasons (Lakhdar et al., 2011). Malignant transformation usually occurs within the primary location of the lesion (Tamura et al., 2006; Lakhdar et al., 2011). Lakhdar et al. (2011) reported the mean age 53 years and female predominance in the cases with malignant transformation of an epidermoid tumour. The mean time to malignant transformation was 14 years (2 to 33 years) (Lakhdar et al., 2011). Tamura and colleagues (2006) reported aged from 36 to 67 years for these patients associated with a mean time of 8.4 years (3 months to 33 years) to malignant transformation.

Rapid development of signs and symptoms is the main clinical feature of malignant transformation, and focal enhancement within the lesion and leptomeningeal metastasis are the main radiologic features indicating the malignant transformation of an epidermoid tumour (Kodama et al., 2007; Lakhdar et al., 2011). Epidermoid tumour is a well-defined lesion that its irregular and nodular surface gives the shiny mother of pearl appearance to this tumour (Chon et al., 2012). This tumour is hypo signal on T1 and hyper signal on T2, and usually without enhancement on GD injection (Tamura et al., 2006). Although in rare cases, it can have minimal rim enhancement (Lakhdar et al., 2011) but appearing of a significant rim or nodular enhancement in epidermoid tumours should alert the physician for a malignant transformation (Kodama et al., 2007).

Malignant transformation can be classified to five groups: 1) primary malignant transformation from an epidermoid cyst, 2) malignant transformation from remnant part of epidermoid tumour, 3) malignant transformation associated with

leptomeningeal carcinomatosis, 4) SCC originated from other benign cysts, and 5) other malignancies from benign cysts (Kim and Kim, 2008).

Malignant transformation in almost all patients occurs at the same site of the primary lesion within surgical field (Fox and South, 1965; Nishiura et al., 1989; Knorr et al., 1991; Murase et al., 1999; Hamlat et al., 2003; Tamura et al., 2006; Kodama et al., 2007; Ge et al., 2009; Hao et al., 2010; Kano et al., 2010; Nakao et al., 2010; Lakhdar et al., 2011; Feng et al., 2014). Tow study reported the malignant transformation in a site adjacent to the primary lesion rather than exactly within the surgical field (Nosaka et al., 1979; Ozutemiz et al., 2017). But only in our study, it occurred within the site away from the primary site and only one case similar to our case has been reported, yet (Chon et al., 2012).

Table 1 summarized the features of some previous studies in this setting.

## Conclusion

Although malignant transformation of an epidermoid tumour is a rare complication, but it can occur. In patients with subtotal resection, rapid and progressive signs and symptoms should be taken seriously. Considering the radiologic finding of malignant transformation, follow-up imaging should be performed either in operated patients or in patients with conservative treatment. Appearing of nodular or rim enhancement in the MRI should warrant for malignant transformation. Our hypothesis is that the malignant transformation can occur away from the primary site of the lesion. But for confirming this hypothesis, it needs more survey of the natural history of the epidermoid tumour. In the setting of occurrence of malignant transformation within the different site from the primary site, complete work up to discover other main probable sites of the tumour including lung and gastrointestinal is mandatory.

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