

## Papillary endothelial hyperplasia of external jugular vein; A rare diagnosis of lateral neck mass

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*Papillary Endothelial Hyperplasia (PEH) is a rare differential diagnosis of a lateral neck mass occur in the head and neck region. A case of Thai male 65 year-old presented with a lateral cord-like neck mass for one week. Imaging studies (USG and CT scan) revealed thrombosis of the external jugular vein with initially unknown etiology. The thrombosed vein was uneventfully excised with a final histopathology report as PEH.*

*This case report described the finding of PEH within the external jugular vein that presented with lateral neck mass, not previously mentioned before in the literature search. Differential diagnosis must be rule out from angiosarcoma due to mimicking pathological character. Complete surgical excision is the treatment of choice.*

**Keywords :** *Papillary Endothelial Hyperplasia, Masson's tumor, External jugular vein, Angiosarcoma, Lateral neck mass.*

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ณปฏล ตังจาดรณศร์ศมี. รายงานผู้ป่วย Papillary Endothelial Hyperplasia ของ External jugular vein ซึ่งมาด้วยอาการก้อนที่ด้านข้างของลำคอ. จุฬาลงกรณ์เวชสาร 2552 ก.ย. - ต.ค.; 53(5): 405 - 11

รายงานผู้ป่วย Papillary endothelial hyperplasia ของ external jugular vein ซึ่งมาพบแพทย์ด้วยก้อนลักษณะเป็นท่อนแข็งที่ด้านข้างของลำคอโดยไม่มีอาการอื่นๆ ผิดปกติ การตรวจทางห้องปฏิบัติการด้วยคลื่นเสียงความถี่สูงและเอกซเรย์คอมพิวเตอร์บ่งชี้ถึง jugular vein thrombosis โดยไม่ทราบสาเหตุ ผู้ป่วยได้รับการผ่าตัด และวินิจฉัยจากการตรวจทางพยาธิวิทยาถึงภาวะดังกล่าว การวินิจฉัยแยกโรคต้องนึกถึง angiosarcoma ไว้ด้วย เนื่องจากความคล้ายคลึงกันของลักษณะทางพยาธิวิทยา การผ่าตัดเอาก้อนออกเป็นวิธีการรักษาหลักในปัจจุบัน

จากการสืบค้นในฐานข้อมูลทางอิเล็กทรอนิกส์ยังไม่พบรายงานรอยโรคลักษณะนี้ในตำแหน่งเดียวกันนี้มาก่อน

**คำสำคัญ :** Papillary Endothelial Hyperplasia, Masson's tumor, External jugular vein, Angiosarcoma, ก้อนที่ด้านข้างของลำคอ

Lateral neck mass had many differential diagnoses and need carefully history taking along with complete physical examination. Papillary endothelial hyperplasia (PEH) is one of a very rare differential diagnosis. It was firstly reported by Pierre Masson,<sup>(1)</sup> "*Hemangioendothelioma vegetant intravasculaire*", in 1923 from hemorrhoid specimen. PEH is also known by many names, e.g., *intravascular angiomatosis*, *Masson's pseudoangiosarcoma* and *Vegetant intravascular hemangioendothelioma*. Clearkin and Enzinger was the first to use this term in 1976.<sup>(2)</sup>

Many PEH were reported with more predilection in head and neck region. Other reported sites include tongue, lips, buccal mucosa, masseter muscle, parotid gland, submandibular gland and also in intracranial (petrous, jugular) region.<sup>(3)</sup> To date six report of PEH<sup>(4)</sup> presented with a neck mass that mainly at anterior neck; include platysma, thyroid, retromandibular space<sup>(5)</sup> and external jugular vein. The case of PEH at external jugular vein previously reported presented with large anterior neck mass.

In this article, the author presents a case of PEH of external jugular vein presenting with a lateral neck mass which has never been reported.

### Search Strategy

In December 2008, an investigation search was performed through the electronic search database (www.scopus.com and www.pubmed.com) on the term: "*papillary endothelial hyperplasia*" or "*Masson's tumor*" with "AND" operator with "*neck vein*".

### Case Report

A Thai male, 65 years-old, who was otherwise healthy, presented with history of 5 days with a slow

growing lateral neck mass on the left side. The mass was first noticed just above the left clavicle with the growth in cranially direction. He gave no history of hematologic abnormality or herbal medicinal usage as well as local trauma and venepuncture on that area. On physical examination, the mass was mildly tender on palpation, cord-like appearance with branching at the level of cricoid cartilage (Fig.1). The mass was extended from the inferior border of left mandible down to the left clavicle without any pulsation. There was no discoloration on the skin. Other otolaryngological examination revealed normal findings. All vital signs are within normal limits. No other mass was detected elsewhere in the body.

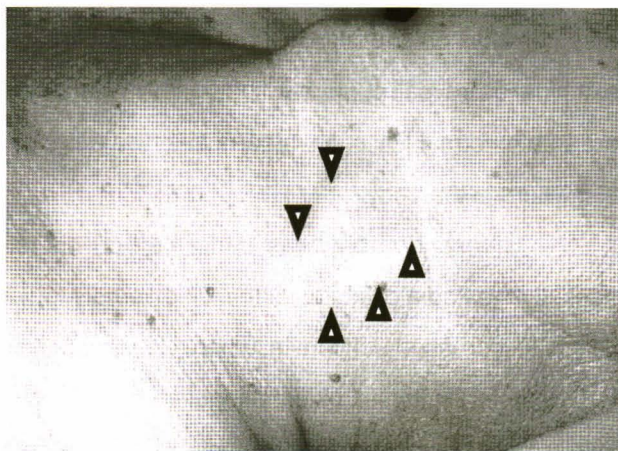
Ultrasonography was performed on his left neck. It revealed a dilatation of the left external jugular vein with increased echo in its lumen. No venous flow was detected in the left external jugular vein. CT scan with contrast was performed and it revealed no enhancement of the left external jugular vein after contrast-administration (Fig. 2). External jugular vein thrombosis was the provisional diagnosis.

The external jugular vein thrombosis, which is an uncommon location, can be occur from many etiologies. Proper investigation more than routine laboratories (CBC) and coagulogram (PT, aPTT) to find out the cause that will help to prevent the thromboses in the future. Due to no extraluminal mass was identified on CT scan, the investigation was performed to search for thrombophilic status more than mechanical obstruction or anatomical abnormality. The screening for thrombophilic included, protein C and protein S level, antithrombin level, also with lupus anticoagulant was done. For acquired thrombophilic status, CA19-9 level with CT scan of chest and abdomen for hidden

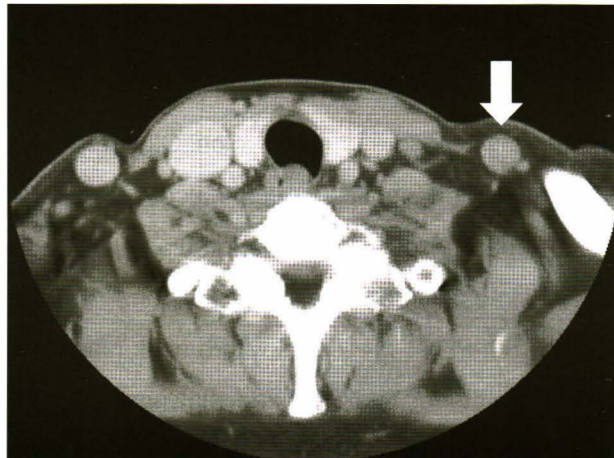
maligancied were ordered. All laboratories value and imaging were within normal limits. (Table 1.)

Surgery was performed to remove the thrombosed vein under general anesthesia with uneventful intraoperative and post operatively (Fig.3 - 4). The patient was safely discharge from hospital with oral anti-coagulant 2 days after the operation. The pathological section shows intravascular lesion consisting of projecting papillary structures

with fibrin cores and bland-looking endothelial lining (Fig.5 - 6). The final diagnosis is papillary endothelial hyperplasia (Masson's tumor) with thrombosis of the external jugular vein. The patient was planned to continue oral anticoagulant and keep INR level around 2 until 6 months post-operatively. The patient came for follow ups at 1-2-4-6 months post-operatively with good condition, no sign and symptom of recurrent disease.



**Figure 1.** Lateral "cord-like" neck mass



**Figure 2.** Post-contrasted CT scan show no enhance of external jugular vein after contrast administration.

**Table 1.** Special hematologic laboratory testing.

Items	Result	Normal Values
Protein C	Function 106%	Function 70 - 140%
Protein S	Free Ag 85%	Free Ag 57 - 158%
Antithrombin	90%	75 - 125%
Lupus anticoagulant	Negative	Negative
CA 19-9	18.97	0-37

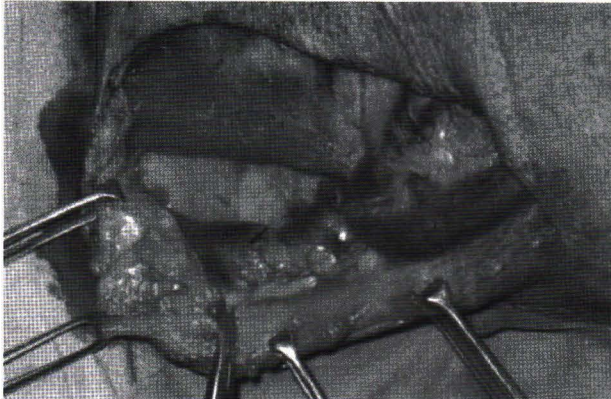


Figure 3. Intraoperative finding of vein thrombosis.

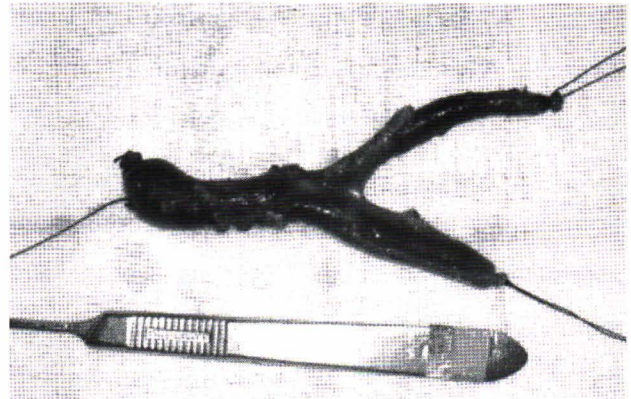


Figure 4. Thrombosed vein specimen with Tumor at stump (Left-handed side).

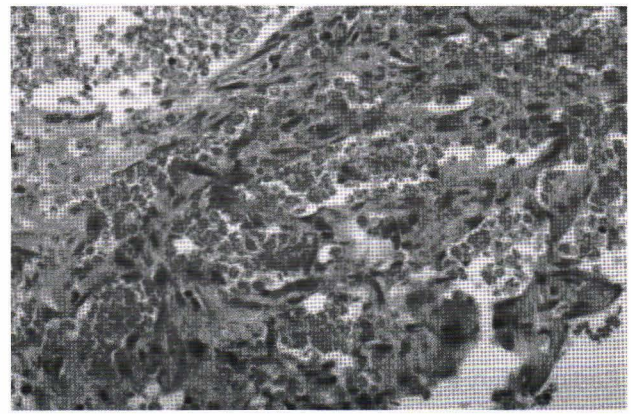
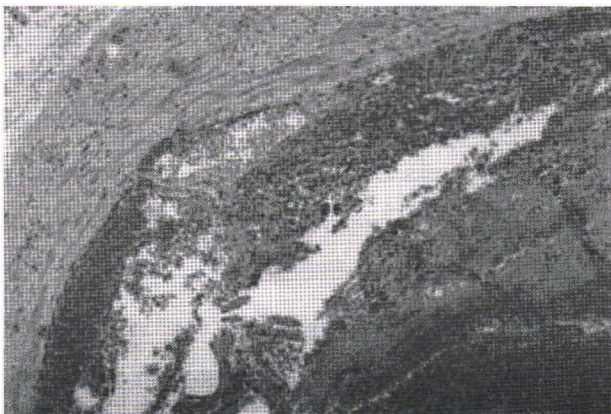


Figure 5. (Upper) and Figure 6. (Right) Pathological section show projecting papillary structure with fibrin.

## Discussion

According to the most articles, PEH can be classified by pathogenesis into three types, namely: <sup>(6, 7)</sup> (1) primary form that newly growth in dilated intravascular spaces; (2) secondary form that occurs in pre-existing vascular lesion (such as AVM , hemangioma, varices); and, (3) PEH arising in hematomas.<sup>(8)</sup> Some physicians believe that the lesion is an unusual form of organizing thrombus. The evidence that PEH originates from organizing thrombus or PEH is a true neoplasm is still unclear. In this patient, the author believes that it was a primary PEH as it showed no other vascular abnormality; hence, it was

identified as pathological and no history of trauma or neck mass like hematoma before.

From literature review, its anatomic localization varies. Pin *et al.* (1993) found that most cases in primary form (56%), followed by secondary form (40%), and in extravascular form (4%). Compared with Hashimoto *et al.*, (1983)<sup>(9)</sup> who reported 33% in primary form, 60% in secondary form and 7% in undetermined type. True incidence report can not be definitely concluded, but extravascular form seems to had the least incidence. PEH also affects females more than males.<sup>(6)</sup> The female:male ratio is 1.2:1.<sup>(10)</sup>

Differential diagnosis is very important because PEH can be mistaken for an angiosarcoma.<sup>(3,7,8,10,11)</sup> Benign features can be identified in PEH, for example, it has no necrotic tissue, low mitosis figures and only slight atypia. It is commonly associated with thrombus.<sup>(8-12)</sup> Immunohistochemistry may be essential for establishment of the vascular nature of PEH, but it does not play any role in differential diagnosis among other vascular tumors.<sup>(3)</sup> Other possible differential diagnosis are pyogenic granuloma and other vascular lesion like AVM, hemangioma or angioendothelioma.<sup>(3,7)</sup>

PEH can be cured by simple excision.<sup>(6)</sup> Recurrence is extremely rare and is thought to occur only when lesion is not excised completely.<sup>(7)</sup> Malignant transformation potential is unclear.<sup>(2,9,10)</sup> Some literature mention about the transformation can occur in lesion that previously incomplete excision. Radiosurgery (g-knife) can be given as main adjuvant therapy in subtotal or partial removal of lesion and may be effective. Chemotherapy has unclear clinical outcome.<sup>(3)</sup> So far, there has been no consensus about the use of oral anticoagulant in PEH. This may simply come from the rarity of this disease.

### Conclusion

PEH can be presented as an isolate lateral neck mass that has no other abnormality. The lesion can be misdiagnosed as angiosarcoma, which is a malignancy and has different management. Complete surgical excision is preferred in resectable areas to avoid recurrence of the disease. Radiosurgery can be given in a partially-removed lesion and it has no definite role of chemotherapy nor oral anticoagulant.

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