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Subdural Hemorrhage in a Child with Klippel Trenaunay Syndrome

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A 10-year-old girl with features of Klippel Trenaunay Syndrome developed a large left frontoparietal subdural hemorrhage. CT angiography and cerebral angiography identified prominent subependymal veins and deep venous system predominantly in the left cerebral hemisphere in association with a dilated left vein of Labbe and hypoplastic superior sagittal sinus. No arteriovenous malformation or fistula was identified. Intracranial hemorrhage in Klippel Trenaunay Syndrome is extremely rare and usually traced to an underlying arteriovenous malformation or fistula. To the best of our knowledge neither a venous source of intracranial bleed nor subdural hemorrhage in Klippel Trenaunay Syndrome has been described in the English literature.

Introduction

Klippel Trenaunay Syndrome is an uncommon mesodermal phakomatosis characterized by the classic triad of cutaneous hemangiomata, venous varicosities, osseous and soft tissue hypertrophy [1]. Occasionally it may result in life threatening bleeding from gastrointestinal tract, genitourinary tract and central nervous system [2]. Intraparenchymal and subarachnoid hemorrhages are the two forms of intracranial hemorrhage reported in Klippel Trenaunay Syndrome [3]. Unlike in other locations, intracranial hemorrhage in Klippel Trenaunay Syndrome has been traced mainly to an underlying arteriovenous malformation [3]. To the best of our knowledge no report exists in the English literature on subdural hemorrhage and a venous source of intracranial bleed in Klippel Trenaunay Syndrome.

Case Report

A 10-year-old female child with left hemihypertrophy and varicosities in the leg with a known clinical diagnosis of Klippel Trenaunay syndrome developed increased somnolence and two episodes of seizure one-week prior and presented to the emergency department. In the past 3 months the child had 3 episodes of trivial injury to the head, which were not associated with headache, vomiting, or loss of consciousness. Her relevant past history was significant for developmental delay and

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Abbreviations: CT, computed tomography; MRI, magnetic resonance imaging

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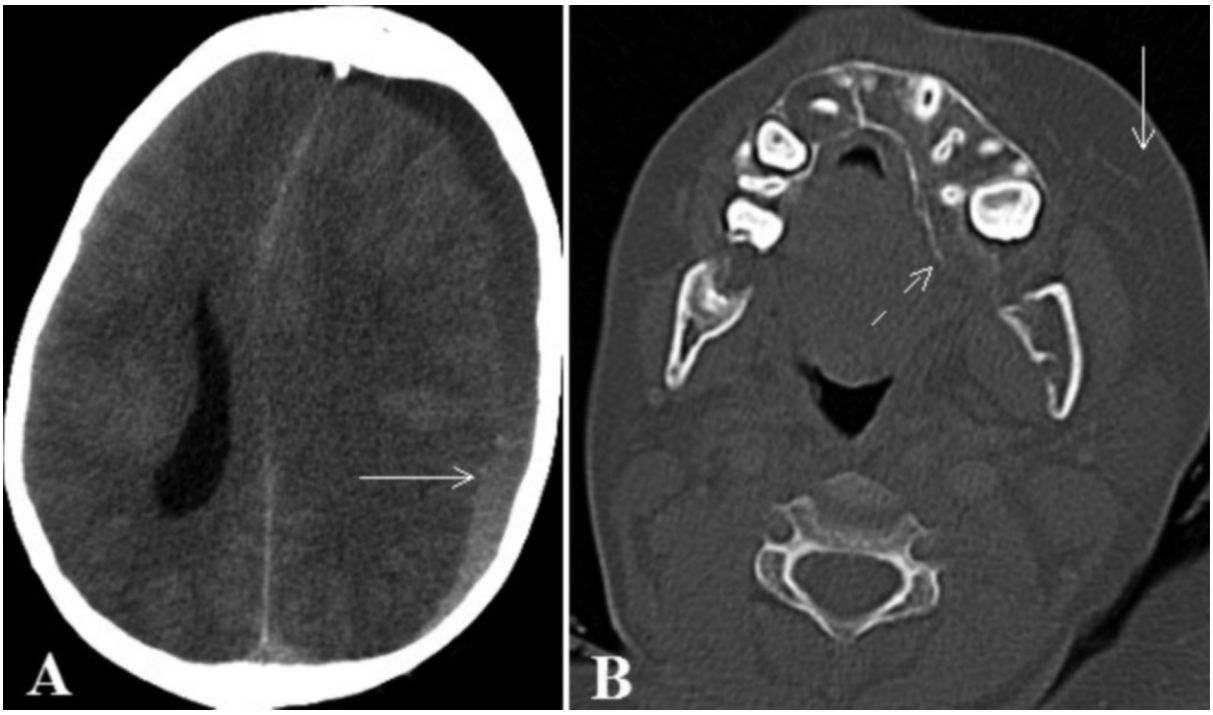

Figure 1. 10-year-old girl with Klippel Trenaunay Syndrome. (A) Non-contrast CT brain shows a left frontoparietal subdural hemorrhage with fluid level causing significant mass effect and midline shift (arrow). (B) Non-contrast CT of the face with bone window shows the hypertrophied mandible (dotted arrow) and subcutaneous tissue of the cheek on the left side (arrow).

refractory seizures. On examination she had a temperature of 37.8 C, heart rate of 117/minute, respiratory rate of 20/minute, and BP of 127/77. The left side of her body showed features of hemihypertrophy. The cardiovascular, respiratory, and abdominal examinations were unremarkable. She responded to verbal query by opening the eyes and spontaneously moved all 4 limbs. The plantars were downgoing bilaterally. A non-contrast CT head showed a large left frontoparietal subdural hematoma with fluid level and significant mid line shift (Fig. 1A). Evidence of left mandibular hypertrophy and soft tissue hypertrophy of the left cheek was noted in the CT scan and MRI (Fig. 1B). CT head obtained 3 months earlier did not show any form of intracranial hemorrhage. Emergent evacuation of the subdural clot was performed. Subsequent enhanced CT head showed multiple prominent transmedullary veins with prominent veins of the deep system, a prominent straight sinus and a left vein of Labbe draining into the prominent left transverse sinus. The superior sagittal sinus appeared hypoplastic (Fig. 1D-F). Cerebral angiography confirmed the venous anomaly. No arteriovenous malformation or aneurysm was noted in the angiography and the arterial phase appeared normal. In the venous phase a prominent left vein of Labbe was noted draining almost the entire left cerebral hemisphere (Fig. 1E). Also there were multiple venous angiomas in the cerebral hemisphere and the cerebellum (Fig. 1F). The cerebral circulation time was within normal limits. Bilateral perisylvian polymicrogyria was observed on MRI brain (Fig. 1C). We also noted hemangioma of the spleen on the CT abdomen obtained 3 months earlier. Following evacuation of subdural hematoma, the patient made an uneventful recovery.

Discussion

Klippel Trenaunay Syndrome is an uncommon mesodermal phakomatosis characterized by cutaneous nevi (typically portwine stain), varicose veins and soft tissue and osseous hypertrophy (4). It was originally described by Klippel and Trenaunay in 1900 [5]. Earlier, Klippel Trenaunay Syndrome was thought to be due to a defect

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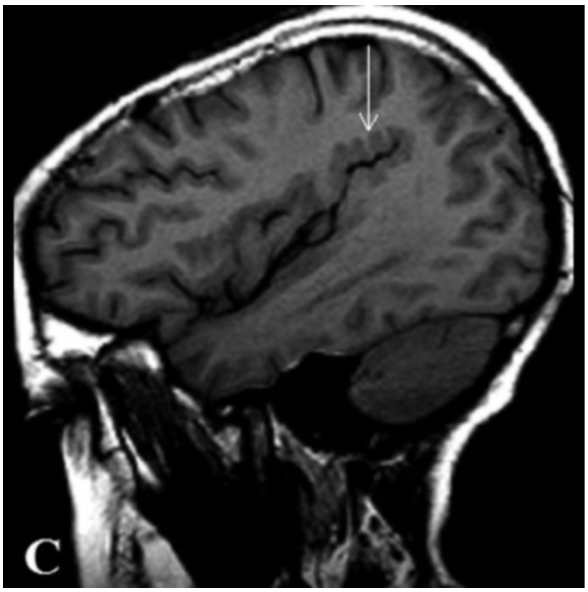

Figure 1. 10-year-old girl with Klippel Trenaunay Syndrome. (C) Sagittal T1 weighted MRI of the brain shows the undulated borders of the perisylvian cortex posteriorly representative of polymicrogyria (arrow).

in the development of the deep venous system of legs. Later a generalized mesodermal defect with particular predilection to the vascular system was favored to explain the multisystem involvement in Klippel Trenaunay Syndrome [6]. Recently genetic abnormalities are increasingly being considered to play the main role in Klippel Trenaunay Syndrome by acting on the angiogenic factors (1). Klippel Trenaunay Syndrome seems to affect mainly the venous component of a vascular tree with manifestations of venous anomalies and varicosities [6]. Involvement of arterial component in Klippel Trenaunay Syndrome is uncommon and may take the form of arteriovenous malformation or arteriovenous fistula. This association is called Klippel-Trenaunay-Weber syndrome since it was Parkes-Weber who originally described this association 7 years after the description of the classic triad by Klippel and Trenaunay [7].

Central nervous system involvement in Klippel Trenaunay Syndrome is considered to be extremely uncommon [3,7]. Arteriovenous malformation and cavernous angioma of brain and spinal cord, intracranial aneurysms, anomalies of the circle of Willis, macrocephaly, microcephaly, hemihypertrophy, hemimegalencephaly, etc. are the central nervous system manifestations reported in Klippel Trenaunay Syndrome [3]. One or more of these findings may explain the developmental delay, seizure, venous hypertension or intracranial hemorrhage occurring in Klippel Trenaunay Syndrome. Intracranial vascular malformations in Klippel Trenaunay Syndrome may cause life-threatening bleeding. Other locations of life threatening bleeding include gastrointestinal and genitourinary tracts [2]. While capillary malformations form the main source of bleeding from gastrointestinal tract and genitourinary tract, in the central nervous system, an underlying arterio-

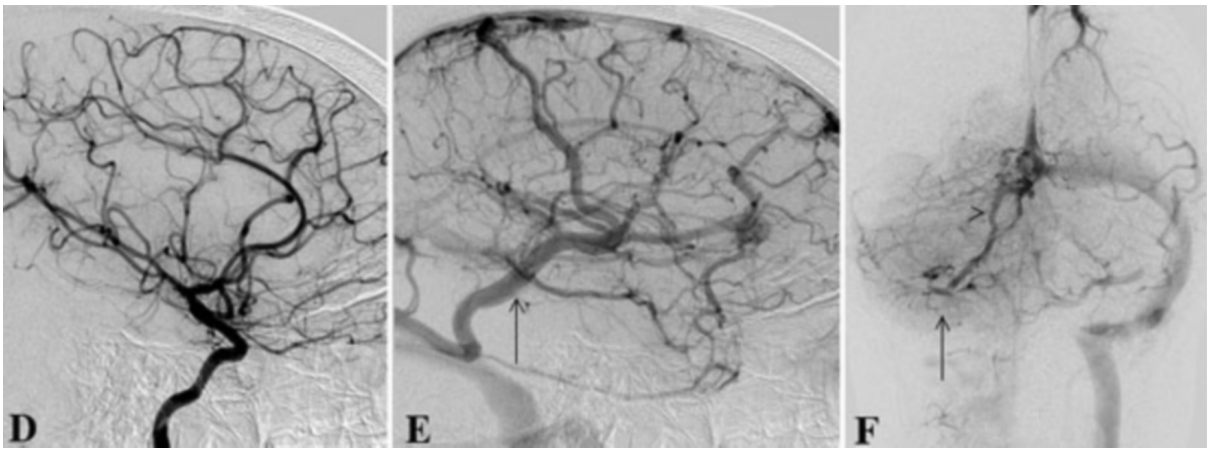

Figure 1. 10-year-old girl with Klippel Trenaunay Syndrome. (D) Arterial phase of the left internal carotid angiogram in lateral view appears normal. (E) Venous phase of the left internal carotid angiogram shows the significantly hypertrophied vein Labbé (arrow) receiving tributaries from a large area. Note the hypoplastic superior sagittal sinus. (F) Venous phase of right vertebral angiogram shows a venous angioma on the right side (arrow) draining through a prominent collector vein (dotted arrow).

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Table 1. Intracranial hemorrhage in Klippel Trenaunay Syndrome

S No	Reference	Age/Sex	Location and type of bleed	Type of vascular malformation	Side of hemihypertrophy
1	Jaksch et al [7]	34 Y/M	Left temporal intracranial hemorrhage	arteriovenous malformation	Right
2	Makiyama et al [8]	8 Y/F	Right parietal intracranial hemorrhage	arteriovenous malformation	Right
3	Taira et al [9]	8 Y/M	Right sylvian fissure subarachnoid hemorrhage	Right middle cerebral artery aneurysm	Right
4	Petzold et al [3]	44 Y/M	Right temporal intracranial hemorrhage	No lesion	Right
5	Our case	10 Y/F	Left frontoparietal subdural hematoma	Venous anomalies in left cerebral hemisphere	Left

venous malformation has been shown to be the source of bleed [2,3]. Our literature search has yielded four reports of intracranial hemorrhage in Klippel Trenaunay Syndrome in the English literature (Table 1). Three of the 4 cases presented with intracerebral hemorrhage and one presented with subarachnoid hemorrhage [7,8]. In 2 of the 3 cases of intracranial hemorrhage, an underlying arteriovenous malformation or arteriovenous fistula was identified as the source of bleed [7,8]. In the third case, cerebral angiogram identified no abnormalities in the intracranial circulation [3]. The case of subarachnoid hemorrhage was related to the rupture of a right middle cerebral artery aneurysm [9].

Some authors believe intracranial bleeding in Klippel Trenaunay Syndrome occurs on the side of subcutaneous and osseous hypertrophy [3]. In 2 out of 3 cases of intracranial hemorrhage, the hemorrhage occurred on the side of subcutaneous and osseous hypertrophy in Klippel Trenaunay Syndrome (7,8). In the case of subarachnoid hemorrhage, hypertrophy was observed on the side of bleed and aneurysm. In our patient also we noted subdural hematoma on the side of hemihypertrophy. It may be possible in Klippel Trenaunay Syndrome, there is a tendency for intracranial vascular malformation and consequently hemorrhage to occur on the side of hypertrophy. Further reports are likely to through more light on this feature. Till then we may have to wait to know the relationship between unilaterality of soft tissue hypertrophy and intracranial vascular abnormality.

Our extensive literature search yielded no report on subdural hemorrhage in Klippel Trenaunay Syndrome in the English literature. It is possible for subdural hemorrhage to occur from an arterial source [10]. However, it is extremely unusual for brain arteriovenous malformations and saccular aneurysms to present with subdural hematoma. Further we did not find an arteriovenous malformation or aneurysm to explain the subdural hematoma in our patient. Almost the entire left cerebral hemisphere was drained through significantly dilated vein of Labbe in our case. It is conceivable the significant venous anomalies noted in our patient on the left side could have contributed to the development of subdural hematoma. Clotting factor deficiency from consumption coagulopathy in Klippel Trenaunay Syndrome may cause intracranial hemorrhage in the absence of an underlying vascular malformation [3]. We did not find a coagulation defect in our patient. The trivial trauma reported in our patient could have been a factor to explain the subdural hematoma but it is highly unlikely to have caused such a large hematoma and fluid level. Makiyama et al reported a lesion similar to vascular nevus covering the dura in their case of Klippel Trenaunay Syndrome with intracranial hemorrhage. However they did not find any connection between the dural lesion and the intracranial vasculature and the intracranial hemorrhage in their case came from the brain arteriovenous malformation [8]. Although visualization of dura was limited by burr holes in our case, it is highly unlikely an undetected capillary nevus of the dura could have been the cause for subdural hemorrhage. We believe the significant venous changes noted over the left cerebral hemisphere must have played the primary role

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in the development of subdural hematoma in our case.

Our case highlights the observation that intracranial hemorrhage can occur in Klippel Trenaunay Syndrome in the absence of an arteriovenous malformation. One should pay attention to the presence of intracranial venous anomalies, which may predispose to the development of bleed. Any neurological event in a case of Klippel Trenaunay Syndrome should draw our attention to the possibility of an underlying vascular malformation and cerebral angiography should be promptly considered without delay to avoid a potentially life-threatening situation.

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