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# Harlequin Syndrome

Anshul Arora\*

Department of Neurology, Fachklinikum Asklepios Lübben Brandenburg, Germany

\*Corresponding author: Anshul Arora, Medical Resident, Department of Neurology, Fachklinikum Asklepios Lübben Brandenburg, Germany, Tel: 004915237846594; E-mail: anshk18@gmail.com

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## Background

A previously healthy 65-year-old woman was referred for evaluation of a peculiar pattern of facial flushing to our hospital.

The symptoms had been present for six months. Relatives had noticed flushing and sweating on the left side of her face following strenuous activity, while the right side remained dry and maintained its normal color. Her medical history included of a right sided mastectomy, Diabetes mellitus type 2 and Hypertension uneventful. For further evaluation she was admitted to our neurological Unit.

## Case Study

On inspection at rest no asymmetric, facial flushing or sweating was noted. Neurological examination revealed no abnormalities Signs of ptosis or miosis could not be noted. The physical examination was eventless.

Vital signs and laboratory values were within normal range as well.

In order to rule out a cerebrovascular insult or perhaps a Wallenberg syndrome a MRI scan of the brain was conducted which showed apart from a generalized microangiopathy no further morphopathological changes.

To assess the autonomic pupillary function a pupillary pharmacological testing was done. The tests of sympathetic integrity included topical Apraclopramide and 0.1% pilocarpine was for the parasympathetic super sensitivity.

The pupils prior and post the application of the eye drops were symmetrical intact.

The patient was asked to run on a treadmill. After 5 min of running at 13 km/h, progressive flushing and profuse sweating could be seen on the left side of her face only.

To exclude a neck dissection and a Pan coast Tumour the patient underwent duplex studies of the carotid arteries and a chest x-ray, each yielding no pathology.

On the basis of the symptoms, we decided to approach it by its fundamentals.

As we recall sweating is regulated by the autonomic nervous which in turn is divided into the sympathetic and parasympathetic nervous systems respectively.

In order to detect an interruption in the sympathetic and parasympathetic innervation in terms of a heart variability rate, a neurophysiological study was conducted.

Interestingly an interruption below the level Th2 could be detected, which not explained the current condition of the patient but also explained the absence of a neurological involvement.

The Patient was informed about the diagnosis and about the benign course of the disease. In spite of having an incurable disorder the patient seemed relieved by knowing the name of her condition.

She was evaluated after 6 months which showed no progression so that further treatment was not required.

## Definition of Harlequin Syndrome

Harlequin Syndrome is a rare condition in the category of the dysautonomic syndrome. It is characterized by asymmetric flushing, neck and sometimes upper thoracic region in response to heat, exercise or emotional factors [1].

Lance et al. first described and named it based on resemblance to colorful harlequin masks [1]. This analogy is also used to describe a severe form of congenital ichthyosis and these entities must not be confused. Occasionally patients with Harlequin syndrome also show other associated autonomic syndromes of the ocular sympathetic as in Horner syndrome/Adie Syndrome/Ross syndrome [2].

## Etiology

The etiologies can be classified into the following: (a) Primary; (b) Idiopathic; (c) Congenital; (d) Secondary; (e) Organic lesion; and (f) Iatrogenic cause.

## Pathophysiology

Understanding the symptoms of the Harlequin syndrome requires an insight in the autonomic nervous system.

The cervicothoracic sympathetic system, supplying vaso-, sudo-, and pupillomotor innervation, acts through a three-neuron pathway.

In the Harlequin syndrome affecting the face the site of the neural sympathetic damage is considered to originate in the T2 or T3 neuron in its course between the stellate ganglion and superior cervical ganglion.

However a lesion proximal to the stellate ganglion can also affect the sudo- and vasomotor innervation to the arm, neck, arm and trunk. As in Harlequin syndrome the extent of clinical signs varies, apparently damage may be very selective. The co-existence of Harlequin syndrome and Horner syndrome suggests pathological lesions of the superior cervical ganglion.

Whereas the combination of Harlequin syndrome and Adie's syndrome implicates a ganglionopathy affecting not merely the sympathetic superior cervical ganglion, but also the parasympathetic ciliary and dorsal root ganglia.

The precise mechanism of axonal damage or degeneration in primary Harlequin syndrome is still unclear.

## Discussion

The Patient presented in this case is a classic example of the Harlequin syndrome they display a primary Harlequin syndrome, idiopathic in origin and associated with a benign natural course.

In this Patient with the help of the neurophysiological study a structural lesion below Th2 as assumed could be detected.

The specific location of the lesion explained the absence of the coexistence of other occulosympathetic symptoms.

In the recent literature the terms Harlequin syndrome and Harlequin sign have been used inter-changeably. However it is preferable to reserve the term Harlequin syndrome for patients that show the paroxysmal signs of hemi-facial flushing.

## Conclusion

Apart from it being less known and less written about the mystery of the patient was able to get solved by approaching it by its fundamentals, making the knowledge of the neuronatomy the most valuable diagnostic test.

## References

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