

An Adult Case of Noonan Syndrome Associated with Persistent Right Primitive Olfactory Artery and Vertebrobasilar Artery Hypoplasia

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Conflict-of-interest statement: The author(s) declare(s) that there is no conflict of interest regarding the publication of this paper.

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Received: May 14, 2018
Revised: June 5, 2018
Accepted: June 8, 2018
Published online: June 27, 2018

ABSTRACT

Noonan syndrome is genetic disease associated with cardiovascular anomalies. Mental retardation is frequently reported in patients with Noonan syndrome; however, reports of intracranial vascular anomalies are rare. Only several cases of Noonan syndrome-associated cerebrovascular disease have been reported. Most of these patients were children or young adults. We examined a nearly asymptomatic adult patient who was diagnosed with Noonan syndrome and found persistence of right primitive olfactory artery and vertebrobasilar artery hypoplasia. This is the first report of persistent primitive olfactory artery associated with Noonan syndrome. Noonan syndrome may be associated with various cerebrovascular diseases, even though there are no

neurological symptoms; thus, the central nervous system and vascular system should be examined in patients with Noonan syndrome.

Key words: Noonan syndrome; Persistent primitive olfactory artery; Vertebrobasilar artery hypoplasia

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Shibata Y. An Adult Case of Noonan Syndrome Associated with Persistent Right Primitive Olfactory Artery and Vertebrobasilar Artery Hypoplasia. *International Journal of Radiology* 2018; **5(1)**: 177-178 Available from: URL: <http://www.ghrnet.org/index.php/ijr/article/view/2318>

INTRODUCTION

Noonan syndrome is genetic disease associated with cardiovascular anomalies. Mental retardation is frequently reported, however, reports of intracranial vascular anomalies are rare. We examined a nearly asymptomatic adult patient who was diagnosed with Noonan syndrome and found persistent right primitive olfactory artery and vertebrobasilar artery hypoplasia.

CASE REPORT

The patient was a 43-year-old man who visited our hospital complaining of chronic mild occipitalgia. He studied at a normal primary and a junior high school, then studied at a high school for disabled children because of mild mental retardation. His surgical history included the closure of an atrial septal defect, the surgical treatment of cockeye and inguinal hernia, and the elongation of his left arm. His height and body weight were 140 cm and 38 kg, respectively. Facial deformities and cockeye were observed; however, no other neurological abnormalities were found. Brain computed tomography (CT) and magnetic resonance imaging (MRI) showed normal findings. Magnetic resonance angiography (MRA) demonstrated persistent right primitive olfactory artery and hypoplasia of the vertebrobasilar arteries and right A1 (Figure). Cervical X-p showed cervical spondylosis.



Figure 1 Magnetic resonance angiography of the case demonstrated persistent right primitive olfactory artery (arrow) and vertebrobasilar artery hypoplasia.

DISCUSSION

Noonan syndrome was first described by Noonan in 1963 and was characterized by Noonan in 1968^[1,2]. The incidence of Noonan syndrome was reported as 1 in 1000-2500 live births. Facial dysmorphism, including hypertelorism, ptosis, downslanting palpebral fissures, low-set ears, short and webbed neck are characteristics of the condition. The responsible gene, protein-tyrosine phosphatase nonreceptor-type 11, has been identified in chromosome 12q24^[2]. Mental retardation and cardiovascular anomalies are frequently observed. Cerebrovascular diseases associated with Noonan syndrome have been rarely reported, including moyamoya disease, intracerebral hemorrhage, cerebral infarction, ruptured aneurysm, arteriovenous malformation and cavernous hemangioma^[3-7]. One patient experienced brain stem and cerebellar infarctions associated with vertebrobasilar arterial hypoplasia^[1]. All of these patients were children or young adults of less than 40 years of age. Our case was 43-year-old man without cerebral infarction or hemorrhage.

Persistent primitive olfactory artery is a rare normal variant; the incidence on CT angiography is reported to be 0.26%^[8], while that on MRA is reported to be 0.14%^[9]. There have been some case reports

of cerebral aneurysm at the origin of a persistent primitive olfactory artery^[10]. This is the first report of persistent primitive olfactory artery associated with Noonan syndrome. Noonan syndrome may be associated with various cerebrovascular diseases; thus, the central nervous system and vascular system should be examined in patients with Noonan syndrome.

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